THE JOURNAL OF NEUROLOGY AND PSYCHOPATHOLOGY

Vol. XIV. JULY, 1933 No. 53

# Original Papers

## THE PATHOGENESIS OF NARCOLEPSY

### WITH A CONSIDERATION OF SLEEP-PARALYSIS AND LOCALIZED SLEEP

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### INTRODUCTION

The purposes of this paper are (1) to call attention to the frequency, in narcolepsy, of a symptom which I propose to call, after Wilson, 'sleep-paralysis'*; (2) to report the case of a patient who had sleep-paralysis and cataplexy; and (3) to show how certain clinical features of narcolepsy may be interpreted in the light of Pavlov's studies of inhibition.

'Cataplexy' denotes transitory paralysis with tonelessness, occurring in response to sudden emotion (usually laughter). By 'narcolepsy' I mean

* The term sleep-paralysis was first used by Kinnier Wilson (p. 90 of his paper), who reported the first two cases in which this phenomenon occurred in association with narcolepsy. The terms predormitium and postdormitium were first used by Weir Mitchell.



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a syndrome characterized by cataplexy and morbid sleepiness (Gélineau's syndrome).

## DEFINITION OF SLEEP-PARALYSIS

By sleep-paralysis I mean paralysis occurring either while the subject is falling asleep (we may call this 'predormitial' or hypnagogic paralysis) or immediately upon his awakening from sleep ('postdormitial' or hypnopompic paralysis). The paralysis may be local or general, in the latter event the subject being utterly unable to move or make a sound. Predormitial paralysis may last as long as several minutes and is succeeded by sleep, save that in some instances the inability to move is associated with anxiety and loss of that peace of mind necessary for slumber, the subject becoming once more fully awake. Postdormitial paralysis, the commoner of the two, lasts from several seconds to several minutes and goes away gradually or suddenly, leaving the subject as normal as ever; in exceptional instances it lasts longer than a few minutes.

Sleep-paralysis was first described by Weir Mitchell who spoke of it as 'night palsy' or 'nocturnal paralysis.' These terms are open to objection, since the phenomenon may occur in relation to diurnal as well as nocturnal sleep, a fact unknown in Mitchell's time. In 1903 Pfister called the phenomenon 'verzögertes psychomotorisches Erwachen.' The phenomenon received little or no further attention until Rosenthal's papers appeared, in 1927 and 1928. Rosenthal retained Pfister's designation of the phenomenon, while terming the individual attacks 'Wachanfälle.' In 1928 Lhermitte wrote of sleep-paralysis, shrewdly terming it 'cataplexie du réveil,' a term whose aptness will presently become apparent.

Mitchell, Pfister, Lhermitte and (in his 1927 and 1928 papers) Rosenthal, were aware of only the postdormitial variety of sleep paralysis. As far as I know, the first instance of *pre*dormitial paralysis was reported in 1928 (Wilson's Case III). In his 1932 paper Rosenthal reported three cases of predormitial paralysis, the attacks of which he designated as 'hypnagog-kataplektische Zustände.'

## THE OCCURRENCE OF SLEEP-PARALYSIS IN CASES OF TYPICAL AND ATYPICAL NARCOLEPSY

Of the more than 200 cases of typical narcolepsy thus far recorded, sleep-paralysis was described in no less than 16. I particularly call attention to this frequency, since no other author has done so, specifically. None of the cases, curiously, was reported before 1928. The 16 cases are the following:—Wilson, 1928, Cases II and III; Brock, 1928; Stransky, 1929, Case I; Zehrer, 1929, Case II; Serejski and Frumkin, 1930, Case I; Redlich, 1931 (an unnumbered case on p. 136); Cave, 1931, Cases V, XVII, XVIII and XXIII; Doyle and Daniels, 1931, Case IV; Rosenthal, 1932, Cases II, III and IV; and Harding and Berg, 1932, Case II.

Thiele and Bernhardt record the fact that of 25 narcoleptics seen at the

Charité Clinic, four had noted the occurrence of 'verzögertes psychomotorisches Erwachen.'

Six more cases have also to be considered. In two of these there were sleep-paralysis and cataplexy, but no morbid somnolence (Rosenthal, 1928, Case VII; Fouché, 1931). In the remaining four there were sleep-paralysis and morbid somnolence, but no cataplexy (Fischl, 1885; Friedmann, 1915, an unnumbered case on p. 92; Rosenthal, 1928, Case V; and Bonhœffer, 1928, Case II).

Riese reported an instance of sleep-paralysis occurring in a patient with cataplexy; we do not know whether the patient also had morbid somnolence.

The following citations illustrate concrete instances of sleep-paralysis.

*Postdormitial Paralysis.*—In Rosenthal's Case V (1928) the patient awoke on two or three occasions 'completely conscious but unable to move. . . . He had a slight feeling of tightness in his chest and a slight feeling of anxiety; finally he "gave a wrench with all his might," . . . whereupon he was able to raise his head and open his eyes, the condition therewith clearing up.'

Wilson thus described the phenomenon in the report of his Case II:—

'Of the greatest interest is the fact that when he has been asleep and dreaming, the emotional content of the dream has precipitated an attack of powerlessness. The worst he has ever experienced lasted for about a quarter of an hour, and occurred under the following circumstances. He had fallen asleep and was dreaming of a murder. The emotion experienced in association therewith brought on one of "loss of power" attacks, so that he at once awoke and was fully conscious but was utterly unable to move a single finger. He thought he heard his brother and sister coming up the stairs; he tried to call to them but could not make a single sound; the more he tried the more intense became his emotion and the more absolute his helplessness; he lay thus, flat on the floor, motionless but suffering acute mental distress, for some fifteen minutes ere the attack dissolved itself spontaneously. Similar but less severe attacks have occurred when he has been dreaming of "terrible happenings" on other occasions.'

As a rule the final 'awakening' from a state of postdormitial paralysis occurs suddenly. Sometimes the merest touch of another person will promptly dissipate the attack—a fact of profound significance, as I shall later try to show. In Brock's case the paralysis generally cleared up by degrees, rather than all at once. To quote Brock: 'Powerless at first, he begins to "wiggle his fingers," then his hands, and then his feet; next he moves a foot and a leg; then he stiffens his abdominal wall; finally, summoning all his strength, he manages to sit up and almost always shakes his head about as if to "clear it." . . . It takes him from five to ten minutes to get up when so seized.'

*Predormitial Paralysis.*—In Rosenthal's Case III (1932) the phenomenon occurred as follows: 'Before falling asleep at night, or when she lies down by



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day, the patient suffers states of anxiety and oppression, as though there were a tight band about her chest ; she is fully awake, hears everything, but can neither speak nor move. People about her notice her condition from her deep breathing.'

Only one patient had both predormitial and postdormitial paralysis (Rosenthal's Case II, 1932).

In two instances sleep-paralysis was incomplete. Zehrer reported that when his patient (Case II) awakes, he ' is fully awake, *can answer*, but for a short time cannot open his eyes or move his body ' (italics mine). In Case II of Harding and Berg postdormitial paralysis was limited to the legs.

Later I shall allude to the frequency and the significance of hallucinations during states of sleep-paralysis.

The following report is of a case recently studied by me.

## PERSONAL CASE

An intelligent, unmarried woman of 21, first seen in July, 1932, complained of a marked inconstant tremor of the upper extremities and the trunk, beginning in 1928.

*Examination.*—The hands, arms and trunk showed an inconstant coarse rhythmic tremor, both at rest and during movement. The family noted that *the tremors were occasionally present during sleep*. There were no alterations of tone. The skin was unusually dry, and since the age of 17 the patient had been abnormally sensitive to cold. However, the basal metabolic rate (August, 1931) was within normal limits. There was evidence of gonadal disorder. The menses (which had begun at 13) were irregular, the periods being separated by as many as six months. Heterosexual interest was weakly developed. Laparotomy disclosed very small hard ovaries.

*Sleep-paralysis.*—In the middle of 1931 the patient began to have the frequent experience of awakening in the morning fully conscious, but unable to move. ' I've tried to lift my arms and legs and turn over, but I just couldn't move. It just seemed like there was no life in the body. I could think of everything and could hear, but the rest of the body seemed to be dead. Then all of a sudden you limber up, and you're able to move again.' The attacks were more apt to occur at the end of a restless sleep. They lasted only a few minutes. She had had no opportunity to learn whether they could be terminated by such extraneous stimuli as a pinch or a loud call.

At times the eyelids were uninvolved in the paralysis, so that the patient opened her eyes but was unable otherwise to move. She never tried to speak.

During these attacks the patient sometimes had auditory and visual hallucinations. Thus she heard her father talking about flowers (they both enjoyed gardening), and sometimes her girl friends. Sometimes, with her eyes open, she saw these people ; they seemed so real that she did not recognize their unreality until the attack had come to a close.

Similar attacks occurred during the night, especially after bad dreams. Thus, when dreaming that a black man was chasing her, she awoke with an attack of paralysis, which lasted several minutes.

Attacks occurred occasionally during the predormitium. ' While falling asleep, I have sort of lost all power for a few minutes, and then I jump—it's a sort of shock, and it wakens me.'

Sometimes the patient awakened able to move, but ' in a daze—I don't know where I am, and I might think it's a couple of days later than it really is.'

*Cataplexy.*—In May, 1932, the patient began to have attacks of cataplexy. ' If anything is real funny, I get sort of weak and shaky in the knees—I get all limber and

drop everything.' On rare occasions the attacks were so severe that she fell: 'my knees get real weak; they sort of slip out.' More often she did not fall, but was required to lean against something in order to avoid doing so. Annoyance sometimes provoked the attacks, as when her younger brothers and sisters disturbed her with their noisy play. Mild attacks sometimes were unassociated with any discoverable emotion. Thus, while reading or writing she occasionally dropped her book or fountain-pen 'for no reason.'

The attacks of cataplexy never lasted more than a minute and there was no disturbance of consciousness.

*Paroxysmal hypertonia.*—In July, 1932, the patient began to have attacks, precipitated by sudden fear, in which she became 'stiff' and was unable to move for several minutes. The significance of this symptom I hope to consider at another time.

The patient at no time in her life showed the least indication of morbid somnolence. Her nocturnal sleep was quite poor and was disturbed by restlessness. She frequently talked and (especially as a child) walked in her sleep.

## THE SIGNIFICANCE OF PAVLOV'S WORK FOR THE MAJOR PHENOMENA OF NARCOLEPSY: MORBID SOMNOLENCE, NOCTURNAL RESTLESSNESS, CATAPLEXY AND SLEEP-PARALYSIS: DEFINITION OF LOCALIZED SLEEP

That Pavlov's work on inhibition is profoundly important for the interpretation of the two cardinal symptoms of narcolepsy (sleep attacks and cataplexy) has already been emphasized by several authors, notably by Adie and Worrall. I shall try to show how Pavlov's work makes possible also the interpretation of sleep-paralysis and of nocturnal restlessness as well as of certain other less prominent clinical features of narcolepsy. It is assumed that the reader is acquainted with the general outline of Pavlov's work and with his concepts of 'internal inhibition,' 'irradiation,' 'concentration,' and 'induction.'

The gist of Pavlov's views on sleep may be stated as follows. Sleep, or inhibition, prevents undue fatigue of the cortical elements, allowing them, after they have been subjected to much stimulation, to recover their normal state. Inhibition is occurring all the time, even in a seemingly alert animal. In the alert animal, however, inhibition exists only in scattered areas of the cortex. When from these areas it irradiates over the entire brain, the animal, as we say, falls asleep. In Pavlov's words, 'internal inhibition during the alert state is nothing but scattered sleep, sleep of separate groups of cellular structures; and sleep itself is nothing but internal inhibition which is widely irradiated, extending over the whole mass of the hemispheres and involving the lower centres of the brain as well. Internal inhibition in the alert state of the animal represents a regional distribution of sleep which is kept within bounds by the antagonistic nervous process of excitation' (p. 253 of his book entitled *Conditioned Reflexes*).

We start out with the assumption that the narcoleptic patient possesses a brain in which inhibition occurs with undue ease. This assumption accords with the fact that the narcoleptic is abnormally susceptible to ordinary sleep-provoking situations. In some cases of narcolepsy sleep attacks occur also in

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response to strong emotions. Here, presumably, the excitatory process associated with the emotion produces, by negative induction, widespread inhibition. In *cataplexy* this induced inhibition is confined to the motor and tone centres, without affecting the ' substrate of consciousness.'

In *sleep-paralysis*, as in cataplexy, there obviously is inhibition of the motor and tone centres but not of the substrate of consciousness. In *post*-dormitial paralysis the inhibitory process existing during sleep fails, as the patient awakens, to disappear from all parts of the brain *simultaneously*. Instead, it disappears from the substrate of consciousness but persists a little while longer in the motor and tone centres, so that the patient awakens to find himself conscious but paralysed. In *pre*dormitial paralysis, as the patient lapses into slumber, the inhibitory process fails to irradiate uniformly over the entire brain. To be accurate, we ought to say inhibitory process*es*, since sleep represents the coalescence of many irradiating processes. In predormitial paralysis these processes invade the motor and tone centres an appreciable length of time before invading the substrate of consciousness.

In certain dogs, under proper experimental conditions, Pavlov found what is virtually an exact duplicate of predormitial paralysis in the human. A remarkable instance is described on p. 266 of his book : ' A dog in which work had hitherto proceeded without any interference by sleep began to show signs of drowsiness—due to its being frequently left in the stand in the experimental room for hours at a stretch without any application of conditioned or any other stimuli. . . . The inhibitory effect of the environment became so strong that the mere introduction of the dog into the experimental room had an immediate and obvious inhibitory effect which became still more pronounced after the animal was placed in the stand. It had to be roused in all manner of ways to keep it from falling fast asleep before the preparations for the experiment had been completed (a matter of only a few minutes). When the experimenter left the room and closed the door in order to start the experiment from outside, and then without losing a minute began to apply one or another conditioned stimulus, the normal conditioned reflex was fully present ; a normal secretion of saliva was obtained and the animal immediately took the food. When, however, after leaving the room an interval was made of four or five minutes before the application of the first stimulus, this stimulus now produced the following remarkable result. The conditioned secretory effect was present and the salivary secretion was sharply augmented on food being presented ; *the animal however did not take the food*, which in order to effect adequate reinforcement had to be placed in its mouth. During this time no relaxation of the skeletal muscles could be observed. When an interval of ten minutes was made after leaving the room no conditioned alimentary reflex could be obtained, and the animal was found fast asleep with a relaxed musculature and occasional snoring. Only one possible explanation of these observations suggests itself. The inhibition must have spread in the first place *only over the motor area of the cortex*, so that excitation

could be initiated by a conditioned stimulus belonging to any other analyser and could spread to the salivary gland but not to the muscles—with a resulting one-sided alimentary reflex lacking its motor component. Later the inhibition spread over the whole mass of the cortex and over the lower parts of the brain, bringing about complete sleep with a relaxation of the skeletal muscles. In this experiment the stages of a gradually developing sleep were brought about . . .' (italics mine). Like a man with predormitial paralysis, this dog in falling asleep passed through a stage in which the centres for voluntary motion were inhibited while the substrate of consciousness continued to be active. This ' demarcation of excitable areas from areas which have undergone complete inhibition ' manifests itself in what Pavlov calls ' localized sleep ' (p. 266).

At this point we may state the conclusion that *cataplexy and sleep-paralysis represent special instances of ' localized sleep,' the motor centres being in a state of dormancy while the substrate of consciousness is in a state of activity.*

Since inhibition sometimes reigns in the motor centres and not the substrate of consciousness, it is of interest to note that it may also reign in the latter and not the former. On forced marches many soldiers sleep. During the sleep attacks of narcolepsy elaborate motor activity is frequently in evidence. Thus many narcoleptics have sleep attacks while walking. In Case II, in my 1929 paper, the patient fell asleep while cycling and continued to ride until awakened by the cries of pedestrians who saw him heading straight for a bus. The patient of Lhermitte and Roques was a stenographer ; during attacks occurring at work she sometimes continued her typing. In these and similar cases, the motor substrate is in a state of activity, while the substrate of consciousness is in a state of inhibition—a type of ' localized sleep ' precisely opposite to cataplexy and sleep-paralysis.

Elaborate motor activity may occur not only during the sleep *attacks* of the narcoleptic, but also during his nocturnal sleep. Contrary to the notion held by some students of the subject, the nocturnal sleep of a strikingly large number of narcoleptics is restless, as has been emphasized by Redlich and Rosenthal. The patients toss about and often talk and walk in their sleep. This point is of the greatest importance, since it prevents the conclusion that the sleep disorder of narcolepsy is simply one of excess. If the sleep disorder consisted only of an excessive need of sleep, we would expect nocturnal sleep to be anything but restless. We must conclude that narcolepsy is characterized not only by an excess of sleep, but also by an *increased tendency to localized sleep.* When inhibition is localized in the motor centres, the patient is conscious but paralysed, as in cataplexy and sleep-paralysis. When it is localized in the substrate of consciousness, the patient is asleep (in the everyday sense of the term), but the motor centres continue to be active, i.e. the sleep is accompanied by an unusual degree of restlessness and elaborate motor behaviour.

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Since there are cases in which motor activity occurs while the subject is fully asleep (using 'asleep' in its ordinary everyday sense), there must a fortiori be cases in which it occurs while he is *partially* asleep. Such cases have been known for many years, especially in the German literature, where they are designated as cases of 'Schlaftrunkenheit' (see, e.g., Gudden's paper). In these cases the subject, having incompletely awakened from sleep, is able to move freely while not yet fully alert.

We shall now consider a number of minor clinical phenomena, which, though they may seem of trivial importance, are really of great significance.

## THE DISPERSION OF LOCALIZED SLEEP THROUGH DISINHIBITION

In Wilson's Case III and Cave's Cases XVIII and XXIII it was reported that the patient instantly recovered from his sleep-paralysis when someone touched him. A similar observation was made in Odell's Case VI; an attack of cataplexy cleared up promptly when the patient was touched. This seemingly strange observation becomes understandable in the light of Pavlov's discovery that inhibition may in numerous ways be dispelled by excitatory stimuli. See, e.g., the experiments on pp. 64 and 65 of his book, showing that an extinguished alimentary reflex may be restored by (*a*) exhibiting a stimulus which evokes an unconditioned defence reflex, or (*b*) combining the conditioned alimentary stimulus with some casual extraneous stimulus. This dispersion of inhibition by an excitatory process Pavlov calls 'disinhibition.' (The nature of disinhibition will be appreciated by anyone who has tried to fall asleep in a noisy room.) In the four cases cited at the head of this paragraph it is evident that the touch of another person dispelled the inhibitory process which at the moment reigned in the motor centres, thereby returning to the patient the voluntary control of his muscles.

It should be remembered that not every excitatory stimulus may cause the termination of an attack of sleep-paralysis. Thus it is nowhere recorded that an attack was terminated by simply shouting at the patient. Moreover, in Wilson's Case III attacks of sleep-paralysis more than once occurred while the patient had a cigarette in his hand, 'and he has actually burnt his fingers badly without being able to make a single movement to let it drop.' Why severe pain should be inadequate, and touch adequate, to terminate an attack is for the present not clear. Since we do not know what part of the cortex is concerned with pain, we cannot discard the possibility that for topographical reasons the excitation of a touch stimulus irradiates to the motor centres more easily than that of a pain stimulus.

In this connection it is pertinent to refer to Cave's Case II, in which the patient by pinching himself brought to a stop his atypical cataplectic attacks, and to his case XXI, in which voluntarily increasing the tension of the muscles of the legs or the tongue served to avert an impending cataplectic

attack. These are two more instances in which excitatory processes, by disinhibition, averted or cut short a manifestation of localized sleep.

In Rosenthal's Case III (1932) it was noted that if during an attack of predormitial paralysis the patient was placed in the sitting position, the attack came to an end. Calling to the patient and touching her had no such effect.

## CLINICAL MANIFESTATIONS OF LOCALIZED SLEEP RESULTING FROM 'CROWDING' OF INHIBITION

The patients in Wilson's Cases II and III, Cave's Cases VI and X, and Rosenthal's Case II (1932) made the observation that attempting to fight off a sleep attack was apt to give rise to an attack of powerlessness. Here again is an observation which at first may seem puzzling, but which is understandable in the light of Pavlov's theory of sleep. Sleep arises through the irradiation and coalescence of many scattered inhibitory processes. When a person fights sleep off, a multitude of excitatory processes come into play, opposing the irradiation of the inhibitory processes. The latter, when not entirely checked, continue to irradiate where they are least opposed. They will invade the motor areas, if these areas are (as in narcolepsy they presumably are) unduly inhibitable.

To my knowledge no observer has made the experiment of forcibly keeping a narcoleptic from succumbing to a sleep attack. It would be desirable to know whether under such conditions powerlessness occurs with significant frequency.

An instructive case is Cohen's Case II. The patient gave the following account of one of her cataplectic attacks: 'I think I could have stopped it in the first stages, but unfortunately I was stupid enough to concentrate not on sitting still, but on trying not to spill the coffee from the cup I was holding, with the result that I kept the coffee cup quite safe, but slipped to the floor beside it.' Here, in the clearest way, we see the 'crowding' of inhibition from one part of the brain to another—in this instance, from the central mechanism of the upper limb to that of the trunk and the lower limbs. One is reminded of the football player who, when rushing the ball, is 'crowded' by his opponents into that part of the field where he meets with the least opposition.

In two carefully studied cases reported by Rothfeld (his Cases II and III) erection habitually followed when the patient attempted to ward off a sleep attack. These patients, also, generally had erections during their sleep attacks. Study of cases of spinal transection has shown that the lower sexual reflexes are inhibited by higher parts of the nervous system. I therefore suggest that in Rothfeld's patients the excessive inhibitability of the narcoleptic brain applied particularly to the higher sexual-inhibitory centres; when sleep was warded off, inhibition was 'crowded' into these centres, their inhibition releasing the lower sexual reflexes.

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### ATTACKS OF POWERLESSNESS 'WITHOUT APPARENT CAUSE'

A number of narcoleptics have reported that in addition to cataplexy (powerlessness precipitated by emotion) they had attacks of powerlessness coming apparently out of a clear sky (Wilson's Cases III and V and Cave's Case XXIII, to mention but a few). I believe that Pavlov's work helps us to understand these attacks. Pavlov has shown that even in the outwardly alert animal inhibitory processes may be demonstrated here and there in the cortex. At just what stage in the irradiation of these processes the individual begins to 'feel sleepy' we, of course, do not know. It may conceivably happen that a sleep attack is 'in the making' and is checked by excitatory stimuli before the patient has had any inkling of the fact that he was on the way to falling asleep. Should the inhibitory processes, checked in their irradiation, invade the motor centres, the patient will be under the impression that he had an attack of powerlessness 'without apparent cause.'

### HALLUCINATIONS AND OTHER DISTURBANCES OF MENTATION IN SLEEP-PARALYSIS

In clinical medicine we expect to find transitional stages between phenomena belonging to the same series. Sleep-paralysis is, in a certain sense, a partial sleep, the substrate of consciousness being 'awake' while the motility substrate is dormant. We therefore ought to find, between sleep and sleep-paralysis, intermediate stages in which the patient is paralysed and consciousness is imperfect. As an actual fact, such intermediate stages are seen in seven cases in which hallucinations occurred during attacks of sleep-paralysis: the cases of Brock; Serejski and Frumkin; Doyle and Daniels; Rosenthal, Cases II, III and IV (1932); and the case which I have here reported.

A discussion of the different levels of mentation would be beyond the scope of this paper. It will suffice to say that the consciousness of a hallucinating person is of an inferior sort. It is the inferior consciousness existing even in healthy people as they gradually fall asleep, when thoughts become 'blurred,' associations follow inferior patterns and hallucinations occur. Thus we may say that in sleep-paralysis accompanied by complete alertness and in the absence of hallucinations, the substrate of consciousness is (as far as we now know) rigidly excluded from the inhibitory process; in sleep-paralysis accompanied by hallucinations the substrate of consciousness is partially inhibited.

In Cave's Cases V and XVIII and in Riese's case, during attacks of sleep-paralysis the patients showed evidence of inferior mentation other than hallucinations.

### THE OCCURRENCE OF POSTDORMITIAL PARALYSIS AFTER FEAR DREAMS

In Wilson's Case II, in Cave's Case XVII and in the case which I have here reported, fear dreams precipitated attacks of sleep-paralysis. The

analogy to the cataplexy of the awake state is obvious. Evidently, strong emotion can not only lead to the inhibition of the (uninhibited) motor centres during the waking state, but, occurring during sleep, can also restrain the disinhibition of the motor centres which is normally incident to awakening.

## SLEEP ATTACKS AND OTHER SYMPTOMS OCCURRING IN CONSEQUENCE OF THE VOLUNTARY SUPPRESSION OF POWERFUL REFLEXES

My second patient, reported in 1929, had made an observation which puzzled me before I became acquainted with Pavlov's work. He stated that on two occasions he had succumbed to a sleep attack in the dentist's chair *while the dentist was actually drilling his teeth*. Considered from the standpoint of everyday experience, sleep precipitated by pain is incomprehensible. But Pavlov has shown us that in this connexion the nature of the stimulus itself is less important than the inhibition associated with it. 'As there is practically no stimulus of whatever strength that cannot, under certain conditions, become subjected to internal inhibition, so also there is none which cannot produce sleep' (p. 252). As an example he mentions an instance in which a powerful electric shock applied to the skin was used as a conditioned alimentary stimulus. After many months of experimentation the stimulus acquired inhibitory properties and became a 'most effectual agent in inducing sleep.' Similarly, in the case of my patient, the painful drilling of the teeth must have acquired inhibitory properties sufficient to induce sleep. In the child painful stimuli arouse uninhibited defence reflexes. As the child grows up a chain of developments occurs, which for the sake of brevity I shall express in the following psychological terms: the child, desiring not to appear a coward, strives to suppress his defence reflex to pain. This process is in all essential respects identical with inhibition. It is therefore understandable that in the dentist's chair pain stimuli arouse powerful inhibitory processes which—in the susceptible person—may eventuate in sleep.

A similar instance has been alluded to by Gowers. The case was that of a man who had attacks of sleep, 'which were induced by most varied influences. He had a nasal fistula, and whenever a probe was passed down this he fell asleep.'

The significant feature of these two cases is that the patients were in situations which required them to bear pain without flinching. If either of them had hurt himself by stubbing his toe, he would not have had to suppress his normal reaction thereto, and would in consequence probably not have fallen asleep.

In this connexion I allude to Rothfeld's Case II. The patient once lay in bed next to a girl and would have had intercourse, had it not been for the presence of a third person. 'He was sexually very excited and suddenly had the feeling as though ejaculation were going to occur. Simultaneously he was overcome by great weakness, so that he lay as though paralysed and

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was able neither to move a limb nor call for help. He was perfectly conscious. This condition lasted a little while, after which he got up and felt perfectly well.' Here the inhibition of a powerful reflex led, not to sleep, but to a form of localized sleep. In principle, this incident duplicates that which occurred in Gowers' patient and in mine.

## CONCLUSIONS

The clinical features of narcolepsy accord with the assumption that the basis of the disorder lies in the undue ' inhibitability ' of one or more parts of the brain. When inhibition invades simultaneously the consciousness substrate and the motility substrate, the resulting sleep attack is indistinguishable from ordinary sleep. The characteristic stamp of narcolepsy lies in the frequency with which inhibition invades one of these substrates without invading the other, thus giving rise to one of the manifestations of ' localized sleep.'

When inhibition is confined to the substrate of consciousness, the patient is ' asleep ' in the ordinary sense of the term, but is restless or performs complex motor acts automatically (narcoleptic patients are frequently restless in their nocturnal sleep). When inhibition is confined to the motility substrate, the patient is conscious but unable to move, as in cataplexy and sleep-paralysis.

An explanation is possible for the following phenomena : the dispersion of cataplexy and sleep-paralysis by extraneous stimuli, such as a touch ; the occurrence of powerlessness in consequence of the effort to ward off an impending sleep attack ; the occurrence of powerlessness ' without apparent cause ' ; the occurrence of hallucinations during sleep-paralysis ; the occurrence of postdormitial paralysis after fear dreams ; and the occurrence of sleep attacks as a reaction to pain that has to be borne without flinching.

It need scarcely be added that a variety of causes may weaken the nervous structures so as to render them abnormally inhibitable, and that narcolepsy is but a symptom of some underlying disease.

## SUGGESTIONS FOR THE CLINICAL STUDY OF CASES OF NARCOLEPSY

I suggest that those who have cases of narcolepsy under their observation pay particular attention to the following questions.

1. In attacks of sleep-paralysis, will a touch cause the attack to go away ? If so, may it be a touch on any part of the body ? What other stimuli, if any, have a like effect ? The same question should be asked in regard to attacks of cataplexy.

2. During attacks of sleep-paralysis and of cataplexy what precisely is the state of consciousness ? Is there evidence of an inferior sort of consciousness, such as hallucinations, dreamy thinking, etc. ?

3. During sleep attacks to what extent is automatic motor activity present ?

4. When the patient tries to fight off a sleep attack, do clinical manifestations of localized sleep supervene? *What happens when the observer forcibly restrains the patient from succumbing to a sleep attack?*

5. What symptoms, if any, appear when the patient is in a situation requiring him to suppress powerful reflexes, as when the dentist is drilling his teeth?

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# RECURRENT POLYNEURITIS IN PREGNANCY AND THE PUERPERIUM AFFECTING THREE MEMBERS OF A FAMILY*

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Recurrent polyneuritis is not a common condition if one excepts cases due to known toxins such as alcohol and lead.

According to Wilfred Harris,[1] Grocco in 1885 was the first to describe recurrent attacks of polyneuritis in the absence of obvious cause. Eichhorst [2] and also Mary Sherwood [3] described two cases from Eichhorst's clinic, each in men; in one both legs were affected, while in the other only the right arm suffered. Targowla's [4] case was that of a single woman who had three attacks of paralysis at the ages of 19, 27, and 39 years; although in many ways similar to those described here (Case III), the attacks were unrelated to pregnancy and the paralysis was more extensive, involving both lower and upper limbs and various cranial nerves. Sorgo [5] recounted the case of a man in whom recurrent polyneuritis was associated with attacks of abdominal pain; and in Thomas' [6] case the subject was a man who suffered also from chronic indigestion. Rabinowitz [7] recorded the occurrence of multiple neuritis in a girl 12 years of age, who had had a similar attack at the age of nine. Holmes [8] mentions three cases observed by himself in one of which the attacks occurred in three successive springs, and cites the seasonal incidence in one of Thomas' cases, where the attacks recurred each June for five years. Bingel [9] recounted the case of a woman of 30, in whom two relapses of polyneuritis occurred, accompanied in each instance by a recurrence of alopecia. Death occurred from respiratory paralysis. Hoestermann [10] described three cases of recurrent polyneuritis, all in adult males, in one of whom thickening of the nerve trunks was noted. Nattrass [11] recorded the occurrence of three attacks of multiple neuritis in a boy in whom various peripheral nerves were greatly hypertrophied. Harris and Newcomb [12] described relapsing interstitial hypertrophic polyneuritis in a man of 52, who died in the sixth attack.

The occurrence of polyneuritis in several members of a family is rare. The progressive hypertrophic interstitial polyneuritis of Dejerine and Sottas [13] was familial in four pairs out of 17 cases reviewed by de Bruyn and Stern.[14] Barker and Estes [15] have described a family hæmatoporphyrinuria sometimes associated with polyneuritis. The two sisters described by Ross

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and Bury [16] appear to be the only recorded instance of *recurrent* polyneuritis affecting more than one member of a family.

Polyneuritis associated with pregnancy or the puerperium has been described, apart from cases due to sepsis or local injury. Farani [17] described a case of polyneuritis beginning in the eighth month of pregnancy, and stated that interruption of pregnancy was sometimes indicated in such circumstances. Albeck [18] recorded four examples in 1926, and later [19] described the symptoms of polyneuritis gravidarum on the basis of nine observed cases. Severe vomiting, a tendency to delirium, marked psychoses, and scanty diuresis appeared to indicate a toxic origin for the polyneuritis. Similar cases have been described by Mills,[20] Ledoux,[21] Bierring,[22] and Bode.[23] In the example recorded by Brugi,[24] on the other hand, there were apparently no signs of severe intoxication. In the case described by Ormerod [25] polyneuritis affected chiefly the flexors of the hips on four occasions, always at the fifth month of pregnancy. Pákozdy [26] recorded the gradual painless onset of paralysis of the fingers and elbows and later of the legs, in a woman five months pregnant. Recovery followed the interruption of pregnancy, but three post-partum relapses occurred, the first three years later, and the others at intervals of 18 months.

According to Harris,[1] puerperal polyneuritis is usually not a painful form, numbness and paræsthesiæ predominating, with weakness and loss of knee-jerks. The same author [27] describes an example of this condition commencing 10 weeks after birth, and in the absence of sepsis. Paralysis affected first the legs, with some sensory loss, then the extensors of the fingers, and later the face and diaphragm, with complete recovery at the end of four months. Batten [28] observed a case in which polyneuritis affecting the hands and the legs with loss of deep reflexes recurred after two confinements. I have been unable to find in the literature any record of polyneuritis, recurrent or otherwise, associated with pregnancy, which has affected more than one member of a family.

The condition described below is of particular interest, in that attacks of polyneuritis of irregular distribution have affected a mother and two daughters at different times over a period of 20 years, and have shown a marked tendency to appear or recur during pregnancy and parturition.

## PERSONAL CASES

*Case* I.—M. W., age 60 years, was healthy until the age of 26 years, when she suffered for about two weeks from ' rheumatic ' pains in the left forearm, without loss of power. She married at the age of 22 and had five pregnancies. The first child was N. M., described below (Case III) ; the second, a female child, died from unknown cause at the age of one year ; the third was T. C. (Case II) ; the fourth is a healthy man, age 25. The fifth pregnancy, 21 years ago, came to full term and a difficult labour terminated with the birth of twins, one living five days and the other five weeks. For three months before the twins were born she suffered very severe pains in the left arm. A few days after parturition she noticed loss of power in this arm, with a complete wrist-drop.

The wrist remained useless for two or three years, and is still weak. There have never been any paræsthesiæ.

Examination on April 4, 1930, showed a partial wrist-drop on the left side, with weakness and slight wasting of the extensors of the wrist and fingers (fig. 1). The left supinator jerk was absent, but there was no sensory loss. No other abnormal signs were found on examination of the nervous system or elsewhere. Skiagrams of the neck showed no evidence of cervical rib.

*Case II.*—T. C., a married woman, age 30 years, was first affected in 1924, when, at the

FIG. 1.—The patient M. W. (Case I) illustrating her inability to dorsiflex the left wrist completely (April, 1930).

age of 20, she complained of pain in the left shoulder, which lasted for a few months, travelled down the arm as far as the elbow, and then went away, leaving no weakness of the limb. Up to this time she had been in good health, except for severe headaches, worse in the morning on waking, and accompanied by dizziness and nausea, which began at the age of 17. She remained well until November, 1926, three months before the birth of her first child, when the pain returned to the left shoulder. The pain began gradually, and worked down to the fingers; it remained as a constant sharp pain, day and night, until the birth of the child. Weakness of the left arm developed gradually during this time. In February, 1927, two nights after parturition, which was normal, she was seized with severe pain in the right arm, and suddenly lost the use of



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the limb. Thereafter the pains continued with diminished intensity, and the power of the arms improved slightly.

When admitted to the Royal Victoria Infirmary on July 1, 1927, there was wasting of both deltoid muscles, the thenar eminences (abductor pollicis brevis) and of the first interosseous space on each side. On the left side there were, in addition, weakness and wasting of the trapezius, supra- and infraspinati, the biceps and the triceps muscles. On faradic stimulation only the right deltoideus and left infraspinatus

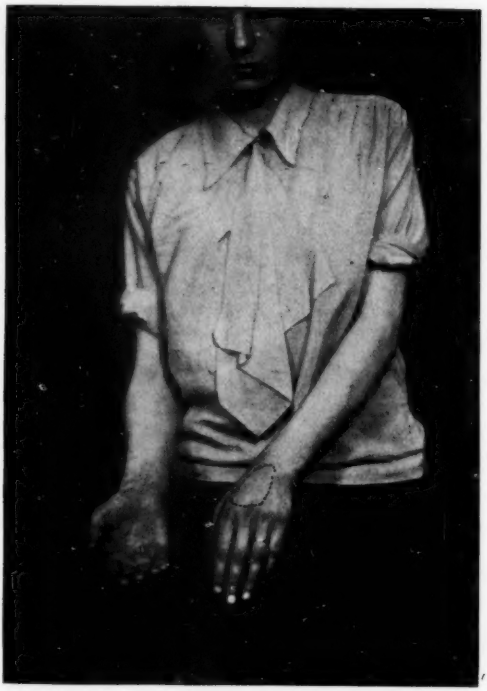

FIG. 2.—The patient T. C. (Case II), showing the area of anæsthesia, enclosed in dotted lines. The right hand has been supinated to show the flattening of the thenar eminence. There is no wrist-drop (April, 1930).

failed to respond. The hand-grip was weak on both sides. The supinator and biceps jerks were absent, but the triceps jerks were active. Sensibility to intermediate degrees of temperature, and to pain, touch and pin-prick was lost over an area along the medial side of both forearms, corresponding to the distribution of the first thoracic nerve. Similar anæsthesia, in this case accompanied by a feeling of numbness, was present over a triangular area on the dorsum of the hands, bounded in front by the roots of the index and middle fingers. Examination of the rest of the nervous system showed no abnormality. No cervical ribs were found on X-ray examination. She was a thin, somewhat sallow woman, but examination of the heart, lungs, abdomen, etc., was negative, and the hæmatological findings were within normal limits. No free

hydrochloric acid was found in the resting juice, but 0·033 per cent. was present after histamine stimulation.

She was discharged from the ward on August 6, 1927, but continued to attend for massage and ultra-violet light treatment for another ten months, during which time there was a gradual increase of power in the limbs.

The patient returned for reexamination in April, 1930. She had remained well for the past two years, except for the area of numbness and hypoæsthesia on the dorsum of the hands (fig. 2). Elsewhere sensation was normal. No muscular wasting remained except for a slight degree in the left supraspinatus and in both thenar eminences, and the power of all muscles was good. The supinator, biceps and triceps jerks were not obtained.

No more was seen of her until August 6, 1932. In June she had had acute appendicitis, for which appendicectomy was performed. The morning headaches, which had occurred almost daily since the birth of her child five years ago, had become worse during the last two years. At times she experienced a little difficulty in raising the arms above the shoulders, and some aching in the deltoid regions. During very cold weather the hands sometimes went 'dead,' the fingers becoming flexed, so that she was unable to straighten them. The area of numbness on the dorsum of the hands remained, and was accompanied by anæsthesia to light touch, and by hypoæsthesia to pin-prick and to moderate degrees of heat and cold. The degree of hypoæsthesia was less on the left than on the right side. The area of anæsthesia to light touch on both hands was greater than at the previous examination, and overlapped the bases of all four fingers. Apart from slight flattening of the right deltoideus, and of both thenar eminences, there was no muscular wasting, and all movements were well performed. Faradic response was good in all muscles.

Her condition at the present time (April, 1933) is unchanged, so that in the absence of further pregnancies she has remained free from relapse for five years.

*Case III.*—N M., a married woman, 38 years of age, had always been healthy except for rheumatic fever at the age of six, and occasional headaches before the menstrual periods.

On October 1, 1929, she complained of pain over the spine of the right scapula, which was severe, constant and worse at night. During the ensuing week the pains travelled to the radial side of the right forearm, and to the dorsum of the wrist and three middle fingers. The fingers began to curl up from weakness of the extensor muscles and the hand-grip weakened. During the second week she developed a complete wrist-drop, with loss of grip and an almost complete inability to move the fingers. The pains then markedly diminished. There was no history to suggest pressure injury, or exposure to cold; she attributed the wrist-drop partly to the fact that at this time she was using the right wrist very much in her work of numbering articles, etc.

During the fourth week (on October 23) she was seen at the out-patient department, where the condition was diagnosed as a musculospiral paralysis due to neuritis or some pressure injury. Massage and movement were commenced, and the wrist supported by a splint.

At the sixth week intermittent pain was felt in the opposite shoulder, again in the region of the spine of the scapula. There were occasional sharp shooting pains in the knees, with some weakness in the legs.

On January 8, 1930, 14 weeks from the onset, pains were still present in the left shoulder and arm. There was no improvement in the power of the right forearm. It was clear that the paralysis was not confined to the distribution of the musculospiral nerve, as she was unable to separate the fingers, or to oppose or adduct the thumb. The supinator jerk was absent on the right side, but the deep reflexes of the upper and lower extremities were otherwise normal. The abdominal reflexes were



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brisk on the right side ; on the left side the reflex was diminished in the upper quadrant and absent in the lower. The plantar reflex was flexor on the right side, and doubtful on the left. There were no other positive findings in the examination of the limbs, and the optic fundi and cranial nerves were normal.

Reaction of degeneration was present in the muscles supplied by the right musculospiral nerve, except in the triceps and supinator longus. A good galvanic response was obtained. Other muscles responded normally to faradism. Skiagrams revealed no cervical ribs and no abnormality of the lower cervical or upper dorsal vertebræ.

FIG. 3.—The patient N. M. (Case III) attempting to dorsiflex both wrists. In addition to drop-wrist on the right side, some flattening of the thenar eminence of the left hand may be seen (April, 1930).

By the end of January there was obvious wasting of the right supinator longus. Early in February the pains increased in the left arm and there developed a feeling of ' pins and needles ' in the ring and little fingers, and later in the middle finger, the paræsthesiæ being increased if she put the arm behind her.

She was admitted to the Royal Victoria Infirmary on February 21, 1930, by which time there had been slight improvement in the grip of the right hand. The bowels were constipated, and the appetite poor, and she had lost a little weight. Menstruation was heavy and occurred every three weeks. Examination showed a thin intelligent woman, of sallow complexion, with no abnormality of the heart, lungs or abdomen. The teeth were artificial, the tongue slightly furred. The optic fundi and cranial nerves

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were normal. There was wasting of the extensor muscles of the right forearm, with wrist-drop (fig. 3). Power in the interossei and adductor pollicis was now normal, and the hand-grip was fair. The left arm showed no loss of power, but some slight wasting of the thenar eminence. The supinator jerk was absent on the right side, but the reflexes of the upper limbs were otherwise normal. There was quantitative diminution of sensitivity to cotton-wool and pin-prick over the ring and little fingers of the left

FIG. 4.—Shows Case III in June, 1932, after the second attack, with wrist-drop on the left side.

hand. On one occasion, when the pain was severe, the patient was unable to feel a pin-prick in this area.

The abdominal reflexes were diminished on the left side. The lower limbs showed normal power, tone, coordination, and no sensory loss. Knee- and ankle-jerks were brisk and equal. A flexor response was easily obtained on stimulation of the sole of the right foot, but the response was less definite and less easily obtained on the left side.

Cerebrospinal fluid obtained at lumbar puncture was normal with regard to its content of protein, chlorides, sugar and cells. A Wassermann test of the blood was negative. No lead could be detected in the urine. The gastric juice contained a plentiful amount of free hydrochloric acid.

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She left hospital on March 25, 1930, but continued to attend for massage and electrical treatment. Pains persisted in the left arm, passing down to the ring and little fingers, for which ultra-violet radiation with a mercury lamp was tried.

With continued massage the right wrist improved so that by the end of May she was able to extend it fully ; the fingers remained flexed, however. Both supraspinatus muscles showed slight wasting, but reacted normally to faradic current.

She married in June, 1930. From July to October her chief complaint was of a tightness in the epigastrium, having no relation to food, and associated with slight

FIG. 5.—Illustrates the winging of the right scapula, the drooping of the shoulder, and the wasting of the trapezius, supra- and infraspinatus muscles (Case III, N. M., in June, 1932).

tenderness and rigidity of the right upper rectus. The bowels remained very constipated.

The upper limbs continued to improve, and massage was discontinued in October, 1930. By February, 1931, she felt absolutely well, and remained so until two weeks after the birth of her first child on February 29, 1932.

Because of the paralysis which her sister had suffered following child-birth, the patient decided to go for her confinement to the Princess Mary Maternity Hospital, where she stayed for 11 days. Four days after returning home pains began in the right shoulder blade, passing down the right arm, as in her former attack. Two days

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later the left shoulder and arm were similarly affected. Subsequently the left leg felt numb and 'dead,' so that she experienced difficulty in walking. Weakness developed in both arms and progressed so that she could not lift the baby, nor raise her arms above her head. The right arm became numb if the weight of the child was allowed to rest upon it.

On April 4, 1932, she still complained of pains in both arms. Examination showed some weakness in the deltoid muscles, with difficulty in raising the arms laterally above the head, and a compensatory rotation of the scapula. All other movements of the upper limbs were normal and there was no wrist-drop. No wasting was present in the muscles of the shoulder girdle, but the thenar eminences showed slight flattening.

FIG. 6.—Shows 'main-en-griffe' on the left side. There is wasting of both deltoids, of the clavicular fibres of the pectoralis major muscles, of the right biceps, and of both thenar eminences (Case III, N. M., in June, 1932).

The remainder of the neurological examination was negative, and no abnormality in the reflexes was found.

She was given a further course of ultra-violet radiation to the shoulders and arms, and told to rest the upper limbs as far as possible until the pains ceased.

A week later she had developed wrist-drop on the left side, and was having severe pains in the left shoulder. The arm reflexes were active and equal. There was diminished sensitivity to touch and pin-prick in the left thumb. Faradic response was obtainable in the paralyzed muscles of the left forearm, but a very strong current was required. The wrist was splinted in a dorsiflexed position.

By May, 1932, the severe pain in the shoulders had disappeared but there was aching in both arms from shoulder to elbow, sometimes passing to the volar surface of the right wrist. She was still unable to dorsiflex the left wrist. Slight wasting of the thenar eminences was present. Massage was recommenced.

On June 20, 1932, examination showed that although the pains had ceased, the

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wasting and weakness of the arms were increasing. Appetite was good, she was not losing weight, and her baby was doing very well. She had difficulty in raising the arms above her head, and wasting was present in the following muscles (figs. 4, 5 and 6): both deltoids, more on the right; the clavicular fibres of the pectoralis major on each side; the right trapezius, supra- and infraspinati, the right scapula being much lower than the left, and the upper angle inclined inwards; the right serratus anterior, with gross winging of the scapula, when the arms were held forward, and to a less extent a similar condition on the left side; the right latissimus dorsi; the right biceps; the left triceps, with inability to extend the elbow completely; the left supinator longus, together with all the muscles on the extensor surface of the left forearm, associated with complete wrist-drop and inability to extend the fingers; both thenar eminences (abductor pollicis brevis), and the left hypothenar eminence. Adduction of the thumb was defective in the left hand. Opposition of fingers and thumb, and movements carried out by the interossei, were not affected. The flexors of the wrists and fingers showed a fair amount of power, but the grip of the left hand was poor. The reaction to faradic current was absent in the following muscles on the left side: serratus magnus, supinator longus, extensor carpi radialis longus and brevis, extensor communis digitorum, extensor pollicis longus and brevis, extensor carpi ulnaris, and abductor pollicis brevis; there was only a very faint faradic response in the extensor indicis. The reaction to galvanic current was delayed, especially in the deltoideus, and was completely absent in the abductor pollicis brevis. On the right side only the serratus magnus failed to respond to faradic current.

Numbness was present over both thenar eminences, but no loss of sensation to light touch or pin-prick could be detected. The skin of the left hand was shiny and atrophic. The deep reflexes were altered as follows: biceps jerk absent on the right side, supinator jerk diminished on the right side and absent on the left. Hoffman's sign and the paradoxical flexor-extensor reflex were negative; and although Wartenberg's sign was a partial positive on the left side the affection of the small muscles of the hand makes it of doubtful value, in this case, as a sign of pyramidal tract involvement. The reflexes of the trunk and lower limbs were normal, and there were no other positive findings in the neurological examination. The movements of the diaphragm, as observed on radiographic examination, were normal.

Her condition at this time bears a striking similarity, in the distribution of muscular wasting, to that found in her sister T. C. in 1927.

From now on there occurred a progressive improvement especially in the power of the right arm. On August 6, 1932, the deltoid muscles showed less wasting, and she was able to raise the arms above the head. Improvement in power and diminution of wasting were noticeable in all the affected muscles, with the exception of the thenar eminences, and of the extensor muscles of the left forearm. There was slight improvement in the ability to extend the fingers, but the wrist-drop remained unchanged. On the right side there was a good response to the faradic current in the serratus magnus, but the rhomboidei reacted poorly, and the biceps gave a negative result with both faradic and galvanic stimulation. On the left side the serratus magnus now reacted neither to faradic nor galvanic current; as on the previous occasion the extensors of the wrist and fingers, with the exception of the abductor pollicis longus, showed no faradic response. Careful testing of the area of numbness on the left thenar eminence showed loss of sensation to light touch (cotton wool), and a diminution to pin-prick and to moderate degrees of heat and cold. A small area of hypoæsthesia to light touch was present over the left deltoid muscle; hypoæsthesia to pin-prick was present in the central part of this area, but heat and cold were correctly distinguished.

When last examined (April 3, 1933) the wrist-drop had almost recovered, the improvement having occurred chiefly during the past five weeks. There was no longer any sensory loss over the thenar eminences or over the left deltoid muscle.

## DISCUSSION

The symptom-complex is that of a recurrent polyneuritis, each attack consisting of three stages :—

1. Severe pains, usually radiating from the spine of the scapula down into one upper limb.

2. The rapid onset of paralysis of lower motor neurone type in groups of muscles of the same limb as the shoulder girdle, sometimes accompanied by sensory loss. The pains remit during this period. Extension of the paralysis, usually accompanied by further pains, may occur during the next few months.

3. Gradual recovery follows during the ensuing 12–18 months, sometimes leaving residual paresis, wasting or sensory loss.

Noteworthy features are the incidence of attacks in three members of a family at long intervals, and the tendency to appear or recur in relation to pregnancy and the puerperium. The paralysis is asymmetrical and frequently widespread, but confined to the upper limbs and shoulder girdles, and not corresponding in its distribution to the areas of supply of peripheral nerves or spinal segments. Sensory loss, when present, is not of glove-and-stocking type, but is confined to small areas of unusual distribution, which may or may not be bilaterally symmetrical, and which may remain after the paralysis has recovered.

Investigations have so far failed to throw any light on the etiology of the condition. The incidence of the disease in three members of a family at different times over a period of 20 years is against the possibility of lead or other extrinsic toxic substance as an etiological factor, nor has there been any evidence of such intoxications. There have been no signs of syphilis or other infection, of pregnancy toxæmia, or of vitamin B deficiency. Examination of the cerebrospinal fluid, gastric contents, blood and urine has revealed no significant abnormality. Hæmatoporphyrinuria has never been a feature of these cases. No cervical ribs and no changes in the vertebral column were visible on radiological examination. There are no nodules to be felt along the peripheral nerves, such as might suggest hypertrophic neuritis.

## SUMMARY

A syndrome is described consisting of lower motor neurone paralysis of irregular distribution, preceded and accompanied by sensory disturbances, which affected three members of a family at different times over a period of 20 years and showed a marked tendency to appear or recur after childbirth.

The mother developed unilateral wrist-drop at the age of 39, a few days after the termination of her fifth pregnancy, having suffered from severe pains in the affected arm for three months beforehand. Similar pains had occurred at the age of 26, but without loss of power. Now, 21 years later, there is some residual paralysis of the extensors of the wrist and fingers. The

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younger daughter, at the age of 23, suffered from similar pains in one shoulder and arm during the latter months of her first pregnancy, with rapid involvement of the other limb after parturition, and subsequently showed in the upper limbs asymmetrical wasting of peculiar distribution, with localized areas of anæsthesia ; gradual but almost complete recovery followed.

The other daughter, at the age of 29, developed severe pains in the right shoulder and arm, followed soon afterwards by wrist-drop, and paralysis of the small muscles of the hand on the affected side. Pains and localized anæsthesia occurred in the opposite arm. Recovery followed and she married. Two weeks after the birth of her first child pains recommenced in both shoulders with paresis of both deltoid muscles. Wrist-drop again developed, this time on the left side, and was followed by extensive but asymmetrical wasting of both shoulder girdles, and of other muscles of the upper limbs. The subsequent recovery has been almost complete.

I am indebted to Dr. George Hall for permission to publish Case III (N. M.), which was admitted to the Royal Victoria Infirmary under his care.

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# FAMILIAL PRESENILE DEMENTIA WITH SPASTIC PARALYSIS *

By

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London

The recognized forms of primary presenile dementia preceded or accompanied by motor disturbances are (1) Pick's disease; (2) Alzheimer's disease; (3) arteriosclerotic presenile dementia; (4) Jakob's spastic pseudosclerosis; and (5) Huntington's chorea.

The chief characteristics of Pick's presenile dementia have been described by Grünthal [1] and Urechia [2] as follows: The condition is most commonly met with in women; the average age is between 50 and 60 years, but it may occur as early as the third decade; the duration of the course is between three and four years, more rarely from two to 10 years; acute and chronic forms have been distinguished; occasionally a family history has been described but no constant etiological factors have yet been discovered. The pathological basis appears to be a primary degeneration of the ganglion-cells of the cerebral cortex, most commonly affecting the frontal lobes and next in order the parietal and temporosphenoidal lobes and less frequently the occipital lobes. The outstanding clinical manifestation of this type is the progressive and eventually profound dementia with considerable disturbance of personality from the outset. Following the earliest stage there is usually no insight displayed by the patient. Most cases are characterized by inertia and lack of initiative together with a progressive global aphasia and in many instances apraxia and agnosia. It differs clinically from the majority of cases of Alzheimer's disease in the frequent absence of restlessness and the extreme rarity of hallucinations and delusions.

In both these conditions there is usually negligible degeneration below the level of the cerebral cortex, whereas in cerebral arteriosclerosis focal symptoms are earlier and more frequent. In the latter condition, further, there is good insight and preservation of the personality until a relatively late stage. Bleuler [3] has said that the main feature of an arteriosclerotic dementia is its lacunar character and that the symptoms have a disturbing effect on the patient, causing considerable anxiety.

The rare condition described by Creutzfeld [4] and Jakob [5] in 1920 and known as Jakob's 'spastic pseudosclerosis' begins at middle age or later

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and consists of slowly developing disturbances of motor and sensory functions ; frequently a tremor of the hands and head is the earliest sign. Gradually the motor symptoms become more pronounced ; slow spontaneous movements, rigidity and tremor are outstanding features. The speech may be slow, monotonous and indistinct. At first the mental symptoms are slight, consisting of anxiety, irritability and depression ; later, confusion, confabulation, hallucinations and finally dementia may appear. A Kayser-Fleischer corneal ring is sometimes observed.

The pathological changes include atrophy of the cerebral cortex and the corpus striatum, especially of the putamen. Histologically, degenerative changes are found in the corpus striatum, thalamus, red nucleus and the dentate nucleus. There is also a diffuse loss in the ganglion-cells with slight increase of glia. The liver may be small, firm and nodular. Jakob, Davison [6] and others have expressed the view that this form of pseudosclerosis, as well as that of Westphal and Strumpell, is closely allied both clinically and anatomically to Wilson's hepatolenticular degeneration.

Huntington's chorea is a definitely inherited malady that usually begins in the fourth decade and is characterized by the development of involuntary jerky movements of wide excursion. Mental changes leading to dementia may accompany or follow after some years the appearance of the choreic movements. In a few instances dementia preceeds the motor disturbances.

## DESCRIPTION OF PERSONAL CASE

C. G., age 47, was first seen on June 14, 1929. He then complained of defective memory, general lack of interest and that people had difficulty in understanding his speech.

*History.*—His speech began to be affected about a year ago and more recently he has appeared less interested and become less efficient at his work, that of engine-driver. He has had no previous illnesses of importance.

*Physical Examination.*—Pupils, optic discs and all cranial nerves are normal. Speech is indistinct and somewhat slurred. Sensation : All forms normal. Upper limbs : Tone of right arm slightly increased ; motor power and all arm-jerks moderate and equal. Lower limbs : Tone generally increased ; knee- and ankle-jerks brisk, right slightly greater than left. Plantar reflexes flexor. Abdominal reflexes moderately brisk and equal ; gait normal. No tremors ; coordination normal. Sphincters intact. Heart not enlarged ; pulse-rate 80, regular ; blood-pressure 155 systolic, 90 diastolic.

By June, 1930, there was no material change in the above condition excepting that the gait had become more deliberate.

*Present Condition :* 1. *Mental State.*—He remains inert all day, sits for hours looking into space and makes no attempt to occupy himself beyond smoking. His mood is complacent and he confabulates very readily. He states that he reads the *Daily Herald* when in fact he never reads at all. He has no idea how long he has been in hospital, stating when asked that he came in last night when indeed he has been in several weeks. He does not know the date or even the month. His remote memory is excellent, e.g., states correctly that he entered the S.E. & C. Railway Service at the age of 13 ; he gives correctly the date of the beginning of the War and also that of the Armistice. He also knows the name of his school and the age at which he left. He

understands all questions readily but has no memory of recent events, e.g., does not know when his wife last came to see him. His speech is scanty, somewhat slurred and indistinct. His attention is fairly good, but there is some delay in answering any question and on some occasions no answer can be obtained. His answers are invariably brief and there is complete absence of spontaneous speech. He is able to repeat a question after one or two minutes and when asked the cause of the delay will say 'I was dreaming.' When up, he goes regularly to the lavatory, but when in bed he is incontinent. He is quite devoid of insight, and retention tests show great impairment. There are no delusions or hallucinations.

2. *Physical Examination.*—Pupils equal, central, circular, react a trifle sluggishly to light but normally on accommodation. Optic discs normal beyond a slight degree of arteriosclerosis. Speech hesitant and somewhat slurred. Overaction of facial muscles and slight tremor of tongue. Sensation normal to all forms of stimuli. Upper limbs: Motor power good but tone definitely increased in both arms. All reflexes, including supinator, biceps, triceps and ulnar jerks, are exaggerated. Lower limbs: Gait slow, deliberate and somewhat spastic on a widened base. Muscular tone of legs much increased, the right more so than the left. Knee- and ankle-jerks exaggerated but equal. No ankle clonus; plantar reflexes flexor; abdominal reflexes not elicited. No tremor of limbs. Coordination of arms good but slight swaying in Romberg position. Arteries only slightly thickened, e.g., radial and dorsalis pedis just palpable. Pulse-rate 76, regular; blood-pressure 140/90. Liver impalpable. Blood Wassermann reaction negative (on three occasions in the last three years). Cerebrospinal fluid: two small lymphocytes per centimetre; total protein 0·05 per cent.; globulin, faint detectable trace: Wassermann and Lange reactions negative.

## FAMILY HISTORY

The accompanying diagram shows the family history as far as it can be ascertained. For the sake of convenience each member is numbered. Our patient, C. G., is no. 29.

His maternal grandmother (4) was born in 1830 and at the age of 40 became afflicted with 'paralysis.' She died at the age of 53. Her sister (5) was affected in exactly the same way at about the same age. Nothing is known of their parents (1) and (2), or of their aunts and uncles. There is said to have been other brothers and sisters but no trace of them can be found.

Our patient's maternal grandmother married into a healthy family, her husband (3) being born in 1829 and dying in 1903 at the age of 74. Until a last short illness he had been perfectly well. They had 10 children. The eldest, Jane (7), was born in 1851 and died at the age of 52. Her husband (6) was a healthy man. The extent of their family is not known but one daughter (24) is known to have died in a mental hospital.

The second son, Charles (8), was born in 1853. He was a coachman by occupation and developed the 'paralysis' at the age of 57; he died at the age of 60. His wife and two children were healthy. The third, Sophie (11), the mother of our patient, was born in 1855. In her case, the 'paralysis' came on at the age of 44, somewhat rapidly, and was followed by insanity. She died in a mental hospital at the age of 54. The records unfortunately

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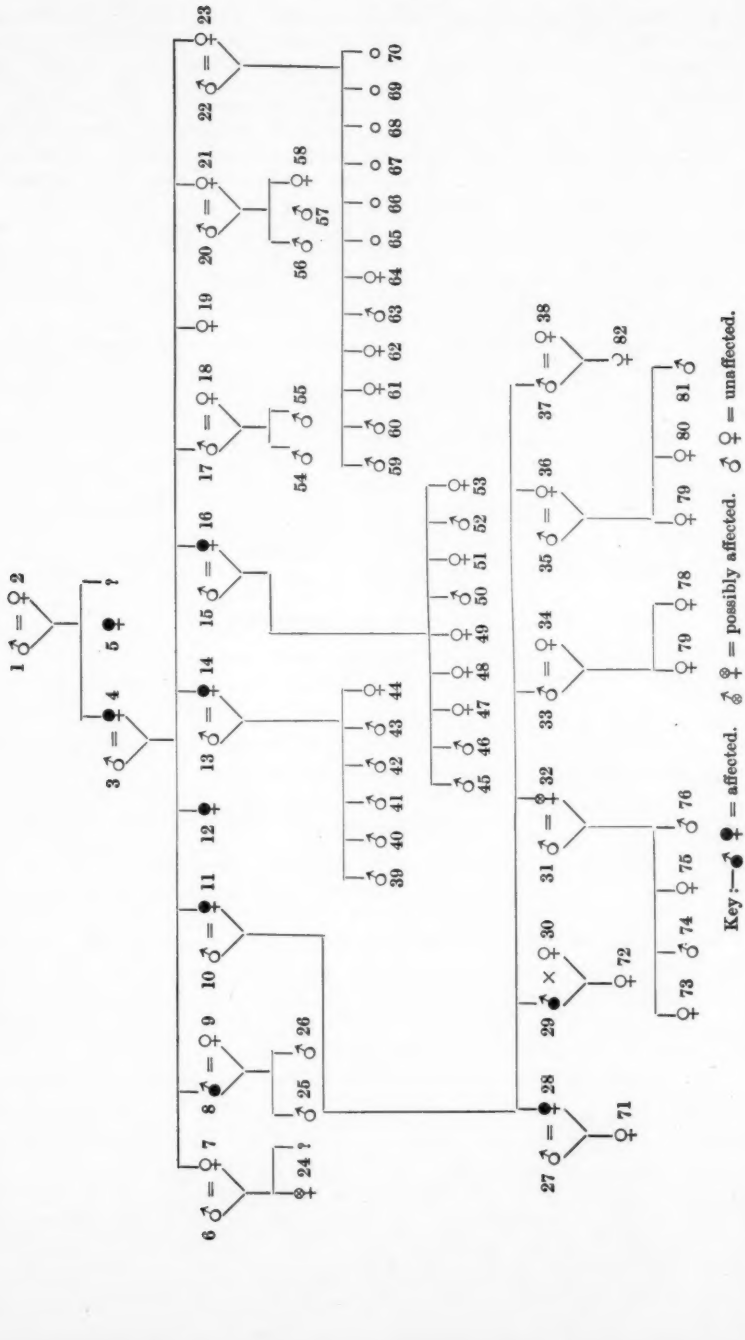
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Key :— = affected. = possibly affected. = unaffected.

cannot be traced. Her husband (10), the father of our patient, came of a healthy family and he himself was well until his terminal illness at the age of 76. Their children numbered six, of whom our patient was the second. They will be referred to later. The fourth, Julia (12), was born in 1857. Insanity began at the age of 48 and she was sent to an asylum. Whilst there she slowly developed ' paralysis ' and died at the age of 53. Inquiry has been made but the asylum notes cannot be traced. The fifth was Alice (14), born in 1861. ' Paralysis ' started at the age of 48 and she died at the age of 66. It was noted that whereas the ' paralysis ' was the same as in the other members of the family, the mental disturbance was less obvious; at no time did she require certification. Her husband (13) was a healthy man and their children (39–44) are all alive and well, with the exception of a daughter who is said to be ' very nervy '; their ages vary from 48 to 29. The sixth was Lorris (16). She was born in 1865, was afflicted with the ' paralysis ' at the age of 48 and died at the age of 50. She was not certified. Her husband (15) and children (45–53) were all healthy. The ages of the children are from 47 to 25. The seventh, Henry (17), was born in 1870. He is now alive and well at the age of 63. His wife (18) and two sons (54–55) are also well; the latter are 10 and nine years old respectively. The eighth was Rose (19), born in 1872. She was mentally deficient, could not walk until she was five years old and died of scarlet fever at 14. The ninth, Ada (21), born 1874, is alive and well. Her children (56–58) are all well, the eldest being aged 36. The youngest of the family, Edith (23), was born in 1875. She had 12 children, six of whom (65–70) died in infancy. The other six (59–64) are alive and well, the eldest being aged 28.

We will now refer to the family of Sophie (11), the mother of our patient. The eldest of her children was Esther (28), born in 1879. She developed ' paralysis ' at the age of 44 and died at 54. We are greatly indebted to Dr. J. W. Wayte of Croydon for a lucid account of the clinical features of this case. He writes that when he first saw her, some two or three years before her death, she presented many features of resemblance to paralysis agitans. Her limbs were held stiffly and she would progress ' by chasing her centre of gravity,' but the facies was never at any time suggestive of Parkinson's disease. After two years her gait changed in character and she seemed to lose her sense of position in space. She now moved stiffly and awkwardly, in so uncertain a manner that it was fascinating to watch where she would finish up her stride. She began to lose flesh and became slowly demented. She seemed to understand much of what was said and tried to answer in a slow scanning manner suggestive of the speech of disseminated sclerosis. She was bedridden in her last year and incontinent of urine and fæces. She became progressively more spastic, lying in bed with knees rigidly drawn up in flexion, but during sleep the rigidities would relax somewhat. She became emaciated towards the end. For many months before her death she would emit spontaneous and apparently quite purposeless cries.

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She would scream for a long time without stopping, but if roused would grin in an understanding manner. She never admitted to having pain. At no time did she suffer from hallucinations or delusions. Her husband (27) was an epileptic and died a few years ago of ' bad health following exposure whilst in war service.' Their only child (71) is 24 years old ; she is of neuropathic disposition and worries unduly about the ' family paralysis.'

Our patient (29) was born in 1882. His wife (30) is a healthy woman and they have one child, a healthy lad of 18.

The third of Sophie's family is Mabel (32), born in 1889. She is now, at the age of 44, under close observation and is believed to be an early case (March, 1933). She herself makes no complaint beyond that for the last two years she has felt continually cold even in warm weather. It is said that all the other affected members of the family had the same symptom a year or two before the appearance of ' paralysis.' Her husband, however, states that she has recently become very irritable and quick-tempered ; ' quite unlike herself,' as he puts it. Also her memory has become faulty ; she will tell her husband or children the same thing at comparatively short intervals. She is neither restless nor emotional but rather apathetic, and has no insight regarding her irritability or her obviously faulty memory. Physical examination shows no abnormality beyond general exaggeration of the deep reflexes.

Her husband (31) is a healthy man and their four children (73–76) are all alive and well, the eldest being 28.

The fourth of Sophie's family is Fred (33), born in 1892. He is quite well, also his wife (34) and two children (77–78), the eldest being 30 years old. The fifth is Violet (36), born in 1895. She is under treatment for anæmia but is otherwise well. Her husband (35) is a healthy man and so are her three children (79–81) ; the eldest is 33. The youngest of Sophie's family is Ernest (37), born in 1897. He is quite well except for astigmatism, and his wife (38) is a healthy woman ; they have a normal son (82), age three years.

It is interesting to note that our patient's maternal grandmother first showed signs of the disease when she was pregnant with her seventh child (17). The eighth child (19) was mentally defective but the ninth and tenth (21 and 22) together with the seventh were considered to be the healthiest of her children in their youth and early adult life. None of them or their families have been affected.

Our patient's mother (11) developed the disease when she was pregnant with her youngest child (37). Thus there are nine cases of the disease spread over three generations (4, 5, 8, 11, 12, 14, 16, 28, and 29) and two possible ones (24 and 32). It is open to objection that many of these cases were in reality not examples of the disease under consideration but more common cases of paralysis such as ordinary hemiplegia. It is, of course, difficult to answer so obvious a criticism, but it is a most significant fact, and one to which much importance may be attached, that the family are fully aware of their ' family paralysis ' and have learnt to recognize its early features.

They are certain that all of the nine members of the family have suffered from the same complaint and they have themselves made the observation that the disease is apparently transmitted through the female side.

Of the members of the family who have died of the disease, the ages of onset were : two at 40, two at 44, three at 48 and one at 57. The longest course was 13 years, the shortest two years and the average eight years. It will also be seen that the tendency is to affect only the elder members of the generation.

## COMMENTARY

The mental changes in the cases we have described resemble those of Pick's disease and of Jakob's pseudosclerosis more closely than those of the other types of presenile dementia. As far as we have been able to trace, however, no such pronounced family history has been described in connection with Pick's disease. A few familial examples have been recorded by Grünthal,[1] e.g., two brothers with the disease ; also in two sisters one of whom had a child similarly affected. Urechia [2] states that heredity plays only a minor part in Pick's disease and that familial cases are very rare. He mentions other types of mental disturbance occurring in close relatives, such as epilepsy, alcoholism and psychoses. Braünmuhl [7] also describes a case of Pick's disease with one brother a general paralytic, one sister psychotic and another sister defective.

Clinically the cases we have described appear to differ from Pick's disease in the early development of spasticity of the limbs and the degree of dysarthria. In our patient there is muscular rigidity of an extrapyramidal type, and, from descriptions, the ' paralysis ' affecting other members of the family involves all four limbs and appears to be of a similar gradually progressive extrapyramidal form. Further, in the majority of cases affected, the development of the ' paralysis ' has preceded that of the mental changes (4, 5, 8, 11, 14, 16 and 28), and in our own patient (29) the first symptom appears to have been dysarthria. In only one case (24) is the mental disturbance known to have preceded the ' paralysis.'

Paresis of both a pyramidal and extrapyramidal type has been described in a few cases of Pick's disease but in all examples it appears to have developed only at a late period. Pathological examination of the brain in Pick's disease, in addition to the degeneration of cortical ganglion-cells, has occasionally shown the cells of the basal ganglia to be affected by the degeneration.

The condition we have described resembles Jakob's pseudosclerosis in the muscular rigidity, dysarthria and mental changes, but it appears to differ from this latter disorder in the absence of tremor and spontaneous movements. In most of the recorded cases of Jakob's pseudosclerosis tremor has been an early and pronounced feature. Also, the inherited factor has not been very striking and there appears nothing in the literature of the condition to compare with the family history we record.

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# THE EXTRACORTICAL MANIFESTATIONS OF CEREBROMACULAR DEGENERATION

By

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## INTRODUCTION

The diseases which form this group were formerly believed to be stereotyped as regards their symptomatology. For instance, Collier and Adie [1] stated in 1926 that 'There are few diseases in which the clinical manifestations are so perfectly regular as this malady.' The sentence was written regarding the congenital or infantile variety, but with the description of the juvenile types by Bielschowsky, Spielmeyer, Vogt and others it is now realized that this group of diseases is no rigid and uniform one, but is an extremely varied and curious collection of syndromes.

The degeneration, if such it be, is not limited to the cortex, though cortical signs and symptoms are characteristic of this amorphous group. Nevertheless, superadded cerebellar signs and symptoms have been described by Marinesco [2] and others. Autopsy reveals extensive degeneration of the cerebellum in such cases. Yet again decerebrate rigidity with concomitant postures, such as flexion of the limbs, labyrinthine and tonic neck reflexes, have been described, notably by Meyer [3] and by Van Bogaert, Sweerts and Bauwens.[4] The causation of such syndromes is presumably the progression of the disease to the caudal end of the red nuclei. Cases do not seem to have been recorded with gross bulbar signs. This is possibly because the involvement of the bulbar structures is incompatible with life. Pathologically, signs of the disease can be found in the cord and even in the peripheral nerves.

With such extensive disease-processes one would hardly expect that cases would not be found which display striatal signs. This is so. In their case Van Bogaert, Sweerts and Bauwens describe a lack of voluntary movements occurring along with the other signs. Again, Marinesco notes that 'Dans la forme tardive, décrite par Bielschowshy, Jansky, Hassin et nous mêmes, il y a des mouvements rythmiques de que même dans ce type on pourrait parler d'une forme hypercinétique'; and again, in describing a case, he says 'En outre le membre supériere droit est animé de mouvements à peu près continuel de caractère choreiforme.' This case showed at autopsy degeneration in the basal nuclei as well as elsewhere. 'Le putamen a un aspect plus clair que le pallidum voisin; cela provient du fait que les cellules pallidales et la névroglie proliférée contiennent plus les lipoides.' And in the globus pallidus 'On y voit l'augmentation périnucleaire du protoplasma microglique, la diminuition des prolongements dépourvés d'épines.'



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With regard to the optic signs it is by no means the rule to observe the cherry red macular degeneration in the juvenile types, but a primary optic atrophy is almost invariable. Naville states that ' Le début est insidieux par faiblesses motrices, cécité et démence progressive, atrophie optique, quelquefois rétinite pigmentaire.' It will be observed in the case recorded below that a generalized pigmented retinitis replaces the usual limited optic signs. That there are transitional types is evident from Goutermann's [5] description that ' The changes at the macula consist of a diffuse pigmentation which extends more or less into the surrounding retina.'

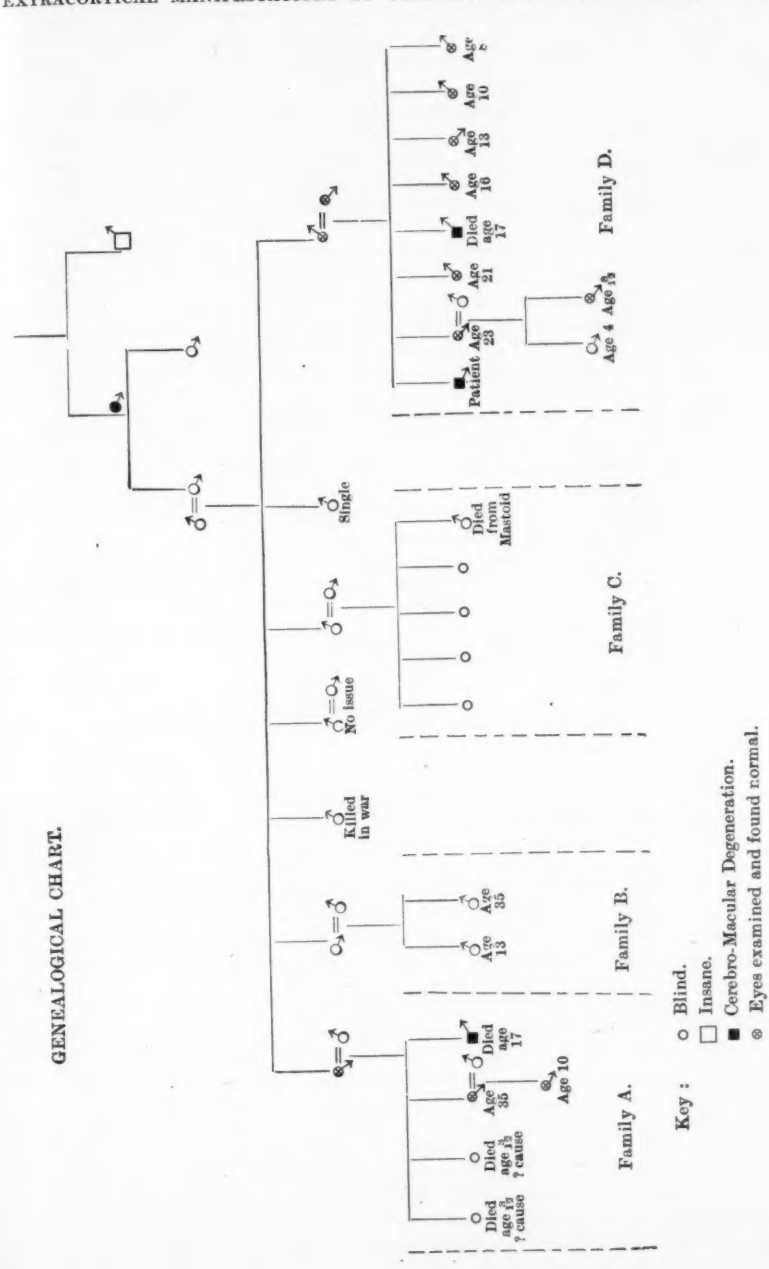
It is difficult to find many references as to the inheritance of the disease. Most writers simply comment on the family incidence. Stenhouse Stewart [6] reports a case in which the disease appeared in a number of the members of two generations. The note is very short, but apparently the case is of the infantile variety. Hansen [16] is of the opinion that a number of factors is needed to produce the typical disease, and ingeniously advances the argument that the abortive, deviating and transitional types are due to a varying admixture of the causative factors. He describes a family in which different types appeared in the different members affected. In Van Bogaert, Sweerts and Bauwen's case the disease appeared in three members of two generations. This certainly does not appear to be suggestive of a Mendelian recessive.

It is interesting to note that in this case (of Van Bogaert) an idiot appeared in the second generation, but lacked the classical signs. Again, in Stenhouse Stewart's [6] case there are recorded eight miscarriages in the marriage of an apparently healthy male member of the family. This occurrence was attributed to the fact that his wife was physically defective. The note does not state in what way she suffered. It should be noted that the mother of one of the cases recorded below had two children who died at the age of three months. It would seem to the writer that these accidents are not altogether unrelated. May it not be that those cases most severely affected are represented by miscarriages, those less affected by deaths in childhood, even without any classical signs ? The idiot would then represent a stationary case wrecked by the disease.

Regarding the pathology there are three schools of thought. (1) It is an agenesis ; for instance, Naville [7] says ' Les faisceaux altérés étaient ceux qui se myelinisent le plus tard et il s'agit d'agénésies et non de dégénéréscences secondaires.' (2) The disease is an abiotrophic degeneration of unknown origin—a widely supported but unhelpful view. (3) The disease is related to Niemann-Pick's [8-15] disease, with which it is sometimes associated, and is therefore a degeneration secondary to the disease in the reticulo-endothelial system. There is nothing in the present case which supports this theory, in view of the fact that the spleen and liver are not palpable.

It is curious to note how resistive the auditory areas are to the degeneration. How frequently there is a true hyperacusis it is difficult to say. The case recorded below shows some diminution to auditory stimuli.

GENEALOGICAL CHART.



## DESCRIPTION OF PERSONAL CASE

HEREDITY.—*Generation* 1.—The writer has succeeded in obtaining an account of the patient's great-grandmother. She is said to have died at the age of 55. She became blind apparently at about the age of 40. She is said never to have been psychotic. Another member of this generation—the patient's great-grand-uncle—is said to have become insane. The writer has tried unsuccessfully to trace this man. He has been assured that there were no signs of blindness in his case. He is said to have died in middle age.

*Generation* 2.—There were two female members of this generation and no males. One of these females died after being married some time. She never had any children. The other female member of this generation is the patient's grandmother. Neither of these women showed signs of blindness or dementia.

*Generation* 3.—This generation is a numerous one. No signs of disease can be found except that the patient's father was born with torticollis. The mother of the patient's cousin (who died of cerebromacular degeneration) is a healthy woman. Neither she nor the patient's father shows any retinal degeneration. For the sake of convenience the patient's mother will be described here. She is a rather stout woman, aged 49. She has a divergent strabismus which she states is due to measles contracted as a child.

The patient's father is aged 53. He was formerly a coal carman, but he has been unable to work for years. He suffers from chronic bronchitis and cannot perform heavy work. Apart from his marked torticollis he displays no obvious abnormality.

*Generation* 4.—There were 17 members of this generation. They are distributed into four families. In two of these families cases of cerebromacular degeneration have occurred. One of these cases is the patient. For purposes of description these families will be called A, B, C and D.

*Family A.*—The two first-born children in this family died at the age of three months. They were never taken to hospital and no definite diagnosis was reached. 'They just wasted away.' A female member of this generation in this family is alive aged 35. She is married, with one child. The fourth member of this family died in a mental hospital at the age of 18. He is said to have been one month premature. He was breast-fed and walked and talked at the age of one year. His mother says he was 'delicate from birth' and he complained of sickness and headaches from the age of two and a half years. He went to school at the age of five and was apparently healthy. His teacher, however, suggested that he had defective eyesight soon after he started school. He was then noticed to be groping for his knife and fork when at table. He 'groped about when in dark passages' (this is very suggestive of nyctalopia). It was also noted that he tilted his head forwards when looking intently at objects. This may have been due to unequal degeneration in the retina with preservation of the lower fields.

He was taken to an Eye Hospital where he was given glasses. His failure to improve led to his being taken to another hospital where the gloomy prophecy of complete blindness in a year was made. He had diurnal enuresis at the age of six and did not appear to be learning at school. He was transferred to a residential blind school, but soon after he went there he had a fit. He had frequent fits following this. He is said to have known when these fits were due and to have been emotional and stubborn before they arrived. He is said to have cried out and then fallen down. He was never hallucinated, but was seclusive and destructive. Owing to the fits he was returned home from the blind school. He was then sent to an infirmary, certified and sent to a mental hospital. There he is said to have become emaciated and not to have known his relatives. He died at the age of 18.

Unfortunately no proper neurological examination was carried out on this patient, neither was any examination made of his eyes. His death was stated to have been caused by status epilepticus.

*Family B.*—This family consists of two males, ages 13 and 25 respectively. They are both said to wear glasses for refractive errors, but are otherwise normal.

*Family C.*—There are five members of this family. One is said to have died from mastoiditis. The others are said to have no signs of illness.

*Family D.*—Out of the eight members of this family two have exhibited cerebromacular degeneration. One died last year and one is the patient.

The one who died last year was a male. He was a full-time birth, born normally, and breast-fed. He walked early and appeared to be a bright child. He was not sent to school until the age of five. A short time after he went to school he was knocked down by a motor car. Following this accident he started to stutter. He was then found to have defective eyesight at school. He was sent to a municipal home for six months. While he was at this home he had two fits. He slowly deteriorated and the number of the fits increased to average one a day. With the increase of the fits his sight gradually failed and he became totally blind. The fits appeared to be generalized and he was dazed after they occurred. This stuporose condition is said to have lasted as long as three hours. He is said to have 'kept pulling his fingers and his teeth' while in this state.

At the age of 12 years he complained of visual hallucinations at night. One night he complained that he saw a rat and complained that he 'saw things' on other occasions. At last he jumped out of bed and declared that there was a dog after him. He was sent to a local infirmary and thence to a mental hospital. He was admitted there at the age of 14 in December, 1928. He was almost blind and had a slight lateral nystagmus, but no other marked nervous signs. In 1931 he had 71 major epileptic fits. He had a pronounced stutter, while some ataxia and doubtful Rombergism were noted. He was seen by the visiting ophthalmologist, who diagnosed cerebromacular degeneration with retinal degenerative chunges.

He died in February, 1932, and no autopsy was allowed.

THE PATIENT.—She was the first-born child of this family. She was a full-time child, normal birth and breast-fed. She seemed rather prone to the usual children's complaints, especially bronchitis. She went to school at the age of three and there she caught measles. She was then kept at home until the age of five when she returned to school. She appeared to be bright and normal until she received a blow on the head at the age of six. It was then discovered that 'she couldn't see to write on the lines at school.' She was taken to a hospital where her eyes were tested and her parents were assured that 'glasses were no good for her.' When she was 12 years old she complained of night terrors but not of hallucinations. She was sent to a residential school, but was returned home at the age of 16, probably because it was found that she could not learn. Even at this time she did not appear to be conspicuously dull and attempted to take part in the domestic work such as scrubbing.

She was kept at home, but at the age of 17 she had her first fit. These fits gradually increased in frequency. According to her mother's statement these fits were preceded by her spinning round and round three times (? epilepsia rotatoria). If not prevented she would fall and hurt herself in

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these seizures. When she fell to the ground she ' had convulsive movements and then seemed dazed for a time.' Her tongue was nearly always bitten. Her fits are said to have been identical with those of her brother. When she was 22 she was sent to an epileptic home and was there until three months ago. She was steadily deteriorating. She was suspected of having a post-encephalitic condition while at the Home, the diagnosis being founded on her ' mask-like face, and tremor of lips and tongue.'

FIG. 1.—Note the attitude of the left hand, the facies and the position of the feet.

Owing to her deterioration she was certified and sent to Bexley Mental Hospital three months ago.

*Physical Findings.*—She is a well-covered woman of 28 years. Her general posture is flexed with bowed shoulders and her hands hanging down before her (see figs. 1 and 2). Her walk is Parkinsonian in type and rather festinate. The nurses state that ' she seems to be tipping forward when she walks.' She does not swing her arms and progresses ' all in one piece ' with no rhythm. There are no tremors of her limbs. She makes no unnecessary

movements and displays little emotion. Her facies is interesting. She has a mask-like expression, but the face itself is full and rounded. The colour is rather high and she usually has a flush. (This fresh-coloured, full but mask-like face was also present in the cousin when he displayed the disease. A photograph which was taken at the age of 13 and which the writer was permitted to inspect showed this very clearly. It is quite distinct from

FIG. 2.—This picture gives an impression of animation which is somewhat deceptive. Note the attitude of the left hand and the bowed stance.

features characteristic of the family—the 'family face' is rather thin and pale.)

*Cranial Nerves.*—Olfactory sense is absent. She was unable to distinguish between oil of lavender and assafœtida. She said that they both 'smelled of nothing.'

She has a very faint perception to a strong electric light if it is shone directly into her eyes. This remnant of her vision is rapidly tired, and

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curiously enough she frequently succeeds in perceiving the light if allowed to use both eyes, but if either eye is covered she says at once that she cannot see it.

My colleague Dr. Walker, who is collecting material for a paper on pigmented retinitis, has kindly allowed me to utilize his notes on this case. He states ' The fundi show large bone-corpuscle-like aggregations of pigment in the periphery and extending to an area of one disc-breadth from the disc. The right macular region is clear, but there are a few masses of pigment in the left macular region. Some of the masses of the pigment in both eyes are long, following the line of the blood vessels and have the " bone-corpuscle " masses branching off from them. The choroidal reflex is otherwise pale. The discs are waxy white. The vessels are thread-like and there is no difference between the width of the arteries and the veins.'

The eye movements are normal. She shows no nystagmus on looking towards her hands. The eyes react to light and give a consensual reflex.

The motor trigeminal shows no abnormality, but she has hypæsthesia all over her face. The conjunctival reflex, however, is present on both sides. Taste is completely absent, and she is unable to tell the difference between salt and quinine She is said to be able to tell the difference between coffee and cocoa, but this is very improbable. There is no perceptible facial weakness. Her hearing appears to be fairly acute. She can hear a watch at the distance of one foot. She hears speech and knows when the gramophone is playing. The ninth nerve appears normal except for the absent sensation of taste over the posterior third of her tongue. The palate moves normally. There is no wasting of the sternomastoids. The tongue protrudes, but not to the full extent. There is a marked tremor. When her tongue is protruded there is a slight tremor in her lips.

*Sensory System.*—She is a poor witness and it is difficult to be sure if her perception to cotton-wool is diminished or not. With pin-prick she feels the prick, but states that it causes her no pain. Although the pin was stuck into her sufficiently to make her bleed she still denied pain. She has no appreciation of heat and cold and constantly insisted that the hot and cold tubes were the writer's fingers. Her joint sense is diminished both for small and large joints. Her sense of vibration is entirely absent. She managed to touch her index fingers together, and to touch her nose, but the performance was clumsy.

*Reflexes.*—Her biceps and triceps reflexes are present and equal on both sides, but the supinator jerks were not obtained (although tested on a number of occasions). The knee-jerks are present and equal, but the ankle-jerks are sluggish. There is no clonus. The abdominal reflexes are present and brisk. The plantars are flexor.

Power seems fairly good. There is no wasting nor trophic lesions. There is a poor peripheral circulation and the legs frequently get blue and cold.

She has cog-wheel rigidity which is variable. At times this rigidity is more that of a vague sense of resistance, but at other times it is marked. The festination of her gait varies similarly.

*Investigation of Other Cortical Functions.*—She is so demented that it is difficult to know how much one can rely on her statements, but the following data are given for what they are worth.

She was unable to identify a matchbox, a thimble, a coin or a pencil by holding them in her hands. She succeeded in identifying a paper, but she may have heard it crackle. She makes no mistakes in words, but she is reluctant to speak and never uses long sentences. It is impossible to test her minutely for aphasia, but this does not appear present with such senses as she has left. She is able to carry out commands such as touching her nose and other parts of her body when requested. She has never learned to knit (an accomplishment usually acquired easily by the blind). This may be due to intellectual difficulties or to slight apraxia. She cannot add two and two together.

*Other Systems.*—She is rather fat and well covered. There is no abnormal hair distribution. Her lungs are normal. There is a soft systolic murmur at the cardiac apex. It is not markedly conducted. Her blood pressure is low, 70/45. The Wassermann is negative in the blood. The C.S.F. was under low pressure—90 mm. of water (when sitting). After about 10 c.c. were removed it refused to register, but could be raised by Queckenstedt's method. The Wassermann and Lange tests were normal. There was no increase in cells or protein.

The liver and spleen are not palpable.

The patient has had three fits in the last quarter. They showed no unusual characteristic. She displays no interest in her surroundings and sits in the same spot all day. She gives no evidence of hallucinations.

In her case the degeneration appears to involve the polar regions of the cortex, sparing the rolandic and temporal areas. The mask-like facies, the tremor of the tongue and lips, the posture, gait and rigidity are all suggestive of extrapyramidal lesions. The hypæsthesia to pin-prick and temperature may be due to thalamic degeneration. It is interesting to note that in Meyer's case considerable changes were found in the thalami.

All the other members of this family are normal. The writer made a special visit to their home (as well as to the home of family A), and although a detailed examination in the neurological sense could hardly be requested the family kindly allowed him to examine their eyes. No signs of pigmented retinitis were found.

*Generation* 5.—One member of this generation belongs to family A. Her eyes appear normal, but there is a little fine pigment on the lateral side of the discs. It probably falls within the bounds of normality.

Of the other two members of generation 5 in family D the writer examined the youngest (æt. eight months) and found no abnormalities in the eye-grounds. The elder member, age four, refused to allow her eyes to be examined.

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## THERAPEUTICS

Apart from preventive measures the writer is unable to find any successful therapeutic attempts mentioned in the literature except those of Vollmer. This experimenter injected an ether-alcoholic extract of brain lipoids and claims that the progression of the disease was arrested. After the cessation of the treatment, he states, the child rapidly deteriorated and died.

A similar experiment, but in this case the peroral administration of raw calf's brain and liver, was made by the same writer. No success was obtained by this method.

These experiments were made on cases of the infantile variety. The writer feels that his case has progressed too far to profit from any therapeutic measure.

He wishes to thank Dr. G. Clarke, Medical Superintendent of Bexley Mental Hospital, and The London County Council for permission to utilize the clinical material; also Dr. F. O. Walker for his kindness in allowing the use of his notes on the eye-condition and of the photographs.

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# CATAPLEXY

By

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The peculiar phenomenon now usually described as cataplexy is no clinical novelty. Its occurrence in association with, or relation to, the syndrome of narcolepsy is well known ; it is sometimes, though erroneously, taken to form an integral part of that syndrome, and is therefore rather apt to be regarded as an abnormal symptom which was unknown until narcolepsy began, as it has of late, to occupy neurological attention more than ever. But while cataplexy as a symptom has features which can be at once detected if it is at all pronounced, minor degrees of the same phenomenon are less easy to recognize, while its physiology and possible biological meaning, as well as its underlying mechanism, are still obscure.

In this brief communication I should like to illustrate the clinical aspects of cataplexy by reference to several fresh cases that have come under my notice, and to make some attempt at understanding its ' meaning ' and at discovering the anatomophysiological basis of its remarkable manifestations.

Some years ago I published a fairly full account of an attack of cataplexy which I had the good fortune to observe from beginning to end as it occurred in a narcoleptic patient ; during that attack I was able to prove that the tendon-reflexes were abolished for the time being, and that an extensor plantar response was present. The essence of the phenomenon was its reversibility ; within a few minutes there was a complete return to the normal status quo ante. One or two further examples may now be cited.

*Case I.*—A male patient, age 36, has suffered from characteristic attacks of narcolepsy for about six years. In addition, he has developed cataplectic fits during the course of the last four years. These are induced by stimuli of an emotional kind, though of diverse qualities.

He had been sitting in my consulting-room for about an hour and was, possibly, rather fatigued, though not, so far as I could see, excited or anxious ; nor had I at that moment been making him laugh. It was about seven o'clock in the evening and he had had a tiring day. However this may be, he suddenly said, ' it's coming on now,' in a slightly indistinct voice, and at the same moment his head fell forward on his breast in a series of little jerking movements. His legs sprawled out in front, but he maintained his seat in the arm-chair, nor did his hands let go of the arms of the chair. There was no alteration of colour, and, as I observed, no particular change in the pulse-rate. Nevertheless he was immobile and speechless for the time. I at once found complete abolition of knee-jerks and ankle-jerks, and on taking off his socks was unable to elicit any plantar response, either flexor or extensor, though his feet were not cold. A few seconds later, he moved his right foot to and fro with a series of slowish movements, limited in range. I asked him what he was doing, but he made no reply. The tonus of the musculature was reduced to one of profound limpness.

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About two minutes later, he shook his head from side to side and muttered in an almost unintelligible voice, that he was ' coming round ' and that the thing was ' passing off.' A moment later he sat up, smiled, and told me in a louder voice that ' it was over.' I now found that his knee-jerks were sluggish, and that his plantars were in feeble flexion. He said, in answer to a question, that he had moved his right foot intentionally, it being the sole part of his whole musculature that he could move. He was inwardly struggling to ' break the spell ' of immobility—in other words, he was in possession of his conscious faculties and was making every effort to innervate his muscles—yet the only outward, visible, sign was a feeble little shaking movement of the right foot.

While this conversation between us was going on, he once more suddenly relapsed into the cataplectic state, and the same phenomena appeared again, just as before. Again there was recovery, and yet again there was relapse. Thus a succession of cataplectic fits—or, at least, a series of partial attacks and partial recoveries—took place after the first one. The time taken from beginning to end, when at length the patient emerged to a normal state without further cataplectic manifestations, was 14 minutes.

The interesting feature of this case, here briefly reported, consists in the occurrence of a *status cataplecticus*, of which, so far as is known to me, no instance has hitherto been put on record. Perhaps the phase might also be described as *serial cataplexy ;* whichever term be preferred, the fact remains that the clinical happening brings cataplexy, by another of its aspects, into line with certain aspects of epilepsy—and this, in my opinion, has its importance.

A second feature is the abolition of the plantar reflex, which would seem to indicate that reflectivity as a whole, both cutaneous and deep, is in abeyance during cataplexy ; and that while the flexor response is absent an extensor response may not take its place.

*Case II.*—A male, age 58, has had characteristic attacks of both narcolepsy and cataplexy for at least 18 years, i.e., since 1914 (incidentally, they began before epidemic encephalitis as known at present made its appearance in England).

During the course of my examination in the consulting-room, without any particular affective stimulus so far as was obvious to me, except that perhaps I had ' stimulated ' him by remarking that I wished I could see an attack and that he was not going to ' oblige ' me, his wife suddenly said, ' He's going to have one now ' ; and as I looked at him his head sank forward and downward, his eyes closed, little flickers appeared round the corners of the mouth. The resemblance to some types of petit mal was very close indeed, but there was no loss of colour, and the body and limbs were immobile. I spoke to him but he made no answer. His knee-jerks, previously active, were at this moment lost. I did not have time to take off his shoes to try the plantar responses, for he lifted his head and said in an indistinct voice that ' there was nothing the matter.' The total duration was but a few seconds, certainly not more than 30 ; and it appeared that all his cataplectic attacks are similarly short.

Of especial interest in this case, aside from the fact that had I not known its nature the superficially close likeness to petit mal might have been misleading, are features which the patient himself narrated in connexion with the cataplectic attacks. In the first place, they are brought on by ' anything

unusual or unexpected,' any experience whose emotional content is definite enough but which varies widely in degree. They were first noticed when he was fishing ; this sport he only took up in adult life, having never fished as a boy. Whenever he hooked a fish he at once ' went helpless,' his legs giving way underneath him, and his grasp on the rod relaxing. So constantly did this—to him unforeseen and inexplicable—phenomenon recur that finally he gave up fishing altogether. The attack, secondly, is accompanied by definite physical sensations : he has a peculiar feeling at the pit of the stomach, ' like a thrill ' ; he has ' a catching at the heart,' sometimes amounting to palpitation ; and, more definite still, it is invariably attended by marked dryness of the mouth and throat—a *xerostomia* which he seeks to overcome by moving tongue and lips, yet the outward expression of this effort, as we have seen, is no more than a few flickers which might pass for those of petit mal.

It is generally admitted that one of the most distinctive characteristics of cataplexy is its occurrence as a sequel to some or other emotional stimulus, of which possibly the most common is one connected with the funny side of things and with the act of laughter. Hence it becomes of interest for our subject to look a little more closely at what actually occurs when laughter grips our sides. There is, first of all, involvement of facial and respiratory musculatures, which is familiar to all ; but if laughter is overwhelming, more escapes from control ; the person concerned ' shakes ' with laughter, he is ' convulsed ' with laughter, he cannot maintain his erect position but may roll on the floor. That is to say, laughter lets loose a series of diffuse, even convulsive movements—and yet the subject is fully conscious, exactly as is the cataplectic despite outward appearances. Again, however, the converse may take place ; instead of being the plaything of movement, the opposite condition affects his frame ; he is ' helpless ' from laughing, he is ' weak ' with laughter, he 'nearly dies' with it. Surely, then, we have here an instance of a double effect, or double possibility—either ' convulsion ' or ' paralysis.' The affective stimulus either excites or inhibits, or inhibition succeeds excitation ; and for some reason at present insufficiently understood, in the case of cataplexy the stimulus seems invariably to inhibit.

Further, the avalanche may extend beyond the skeletal to the nonstriped musculature ; visceral accompaniments are obvious enough in the case of laughter—the skin glows and reddens, the eyes sparkle, increased glandular secretion occurs. Analogous or similar phenomena may be in evidence when the subject is seized with weeping as the result of an appropriate emotional stimulus ; his body is ' shaken ' with sobs, he too is ' weak ' from the experience, and in certain cases the escape involves even decontrol of visceral apparatus, just as in cases of excessive laughter.

Brissaud recounts the history of one of his patients which is apposite in this connexion.

' Une dame lui annonce que son petit chien vient de mourir. " Vraiment (dit-il) ; oh ! que c'est regrettable ! " Est-ce le dernier mot " regrettable " qui, par une sorte

de suggestion, réveille en lui une série d'idées tristes ? Toujours est-il qu'il se produit comme un déclenchement de tous les ressorts expressifs ; la simple tristesse de la physionomie entraîne les larmes ; après les larmes, les sanglots ; bien plus, ce malheureux homme n'est plus maître de ses sphincters.'

Thus at the bidding of imperious emotion released mechanisms come into action, and both bladder and rectum may void their content. That such occurrences are not confined to persons actually suffering from some nervous affection but may be developed in others who are ' normal,' there is abundant testimony from the colloquial speech of different races of mankind. The significant point which emerges for our discussion is, that there must be radiation of the original affective impulse from an emotional ' centre ' to a number of somatic and visceral ' centres ' in physiological relation therewith.

At this point it becomes desirable to note that varieties of the same series of events are to be found in some patients who also suffer from epilepsy, particularly if that epilepsy is conditioned by stimuli of a psychical—that is to say, affective—kind.

*Case III.*—A male patient, age 28, has suffered from epileptic fits, both major and minor, since he was 14 years old. The major attacks, which occur but rarely, are always preceded by a vivid dream, of an unusually terrifying sort ; he imagines in his dream that he is about to be hanged, or shot, for some misdemeanour—sometimes both hanged and shot (!) ; the origin of this nightmare has been successfully traced but need not here be detailed. Sometimes the dream antedates the fit by 24 hours, sometimes much less, but the fit itself only occurs in sleep, even sleep of a few minutes after a midday meal or for any other reason.

The minor attacks are so brief as to pass almost unnoticed, and do not appear to be activated by any discoverable stimulus.

In addition, however, the patient has observed for years that under any circumstances of excitement, pleasant or unpleasant, under the influence of unusual emotional events, he always loses control of his bowel. So much has this been the case that it has been regarded as a sort of ' family joke.' When out hunting rats with a terrier, when shooting pheasants, he has had repeatedly to ' retire ' in view of the imminent evacuation of the rectal contents. This has sometimes happened three times of a morning, when engaged in sport. Similarly, when once called upon to make a speech unexpectedly at a social gathering, he had to leave the room.

The clinical features of this peculiar case are very instructive ; an emotional experience reverberates in his dreams and precipitates his epilepsy ; affective excitants also cause decontrol of visceral mechanisms. On the other hand, however, he has never had any attack recalling those of full-blown cataplexy.

Evidence which need not here be recapitulated has been gradually collecting in favour of the belief that the optic thalamus, in particular the palæothalamus, is related to afferent impulses laden with emotional tone ; and that the expression of the emotions, in movement of skeletal muscle and activation of neurosympathetic mechanisms, is initiated therefrom. To exclude the cortex however from the succession of events is impossible, for the impulses, coloured with affect, are appreciated in consciousness and have

therefore presumably reached the percipient cortex. Thalamofugal impulses are capable of acting on the cortex, in a plus or minus direction. They stimulate or inhibit, according to circumstances. One of the proofs of this is, that in cataplexy the patient may be unable to speak. No doubt the muscles of phonation and articulation are temporarily paralyzed in cataplexy, just like other musculatures ; but when a person is ' speechless ' he is something more than ' inarticulate.' The cataplectic may be ' deprived of speech ' though the attack has not radiated through the whole body.

*Case IV.*—A case in point is that of a narcoleptic and cataplectic subject who once saw approaching him on a thronged street a friend whom he had not seen for many years. At the very moment of pleasant anticipation, just when he was walking forward to greet him, the emotion of the moment overpowered him ; ' rooted to the spot,' he could not hold out his hand, could not say a single word ; the friend passed without observing him and was lost in the crowd ; by the time my patient had recovered, he had disappeared.

It is, I submit, plausible that inhibition of the cortex had taken place as a result of stimuli reaching it via the thalamus. But we must hasten also to note that this postulated inhibition is local and restricted. For observation and the statements of our patients convince us that during the average fit of cataplexy consciousness is preserved and therefore the cortex as a whole has not undergone abrogation of its functions. There is a block somewhere between ' volition ' and ' innervation '—just as there is in hysteria. Where precisely this occurs, in an anatomophysiological sense, is peculiarly difficult to determine ; it appears to me more feasible to suggest that it occurs at the middle (rolandic) level than anywhere else. We know that the ' will to innervate ' is maintained, for sometimes cataplectic patients say that they ' struggle inwardly ' to move, though nothing is effected.

Yet the fact must not be lost sight of, that the musculature is, for the time being, emptied of tone, and that the reflexes are in abeyance. Ordinarily, when lesions of the upper motor neurones develop slowly, lower automatisms come into increased activity, so that the reflexes are exaggerated and involuntary spasmodic movements may occur. But in the case of sudden shock-like lesions of the upper neurones, the lower neurones are simultaneously affected, and reflectivity may be entirely lost. Hence the flaccidity and the reflex loss of the limbs cannot—in the case of cataplexy, which is as fulminant as any shock—be taken by itself to prove that the site of the physiological disorder is at a low (spinal) level. The truth may well be, that instantaneous inhibition radiates through upper and lower motor neurones alike, universally.

It is striking to think that this can and does occur with such rapidity because of emotional experience, by no means always profound—sometimes trifling or almost unrecognizable ; but it supplies an excellent illustration of the reaction of the organism *as a whole* to affective stimuli.

If, then, we may suppose that the motor characteristics of cataplexy are the visible expression of total and immediate paralysis of the skeletal



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musculature by inhibition of its nerve-cells of origin—a single massive motor unit—a ' motor ganglion,' acted on powerfully by sensory excitation, we must also seek to ascertain whether the hypothesis will account for the atony of the muscles. That tonus must be represented at the cortical level has long been considered by myself a rational inference from various clinical data at our disposal ; and at the Berne Congress of 1931 I made a brief communication urging the need to accept this view if these clinical data are to be reasonably explained. At the same Congress Sir Charles Sherrington tentatively expressed the conclusion to which he was inclined to turn, viz., that tonus and movement are one and the same. ' Tone follows movement like a shadow.' Should this theory receive support, it will explain the motor phenomena of cataplexy without much difficulty ; for we should then affirm that inhibition of the unified ' motor ganglion ' from the sensory side simultaneously entails inhibition of tonus throughout the musculature innervated from that cortical ' ganglion.'

In addition, however, to thalamofugal action on the motor cortex, thalamohypothalamic radiation is equally characteristic of the cataplectic attack. Immediately preceding the latter, or with it, symptoms occur which are referable to visceral centres situated round the ventricular system, reaching from the floor and sides of the third to the floor of the fourth ; the patient ' loses his breath,' feels ' hot ' or ' cold,' has a sensation at the pit of the stomach, his heart palpitates, and so forth. During the fit itself there may be pallor, perspiration, etc. The xerostomia of one of my cases is also classifiable in this neurovisceral series (it is interesting to note that the symptom has been observed in thalamic lesions apart altogether from its appearing in connexion with cataplexy). Although the main incidence of the attack is on skeletal musculature, these visceral concomitants merit attention, since they indicate spread downwards as well. Further proof of this is provided by the observed fact that sometimes cataplectic and narcoleptic seizures are interchangeable, sleep taking the place of the motor phenomena, and there is good reason to believe that sleep is related to the function of some periventricular ' centre.'

The emotion which is so profound as to cause relaxation of sphincters does not seem to characterize cataplexy ; nevertheless we have the record of one of Redlich's cases, in which hearty laughing led to an attack of powerlessness which culminated in loss of both vesical and rectal control. Extensive radiation of the kind, however unusual, may therefore be found on occasion ; and in this respect we find an epileptic analogy worthy of note.

The ' storm centre ' of the cataplectic seizure would thus appear to be thalamic, with immediate spread to cortex in part and to hypothalamus and ventricular floors in part.

It is desirable, in conclusion, to take a still wider view of the phenomena under discussion. Have they a biological meaning ? To quote from a previous article of my own : ' Is it conceivable that we have here through disease

stumbled upon a biologically old automatism, a defence-reaction in which immobility is of advantage to the frightened animal, " struck all of a heap " ? Is cataplexy akin to the Totstellreflex, where the animal is " frightened out of its life " ? ' The cartoonist of the War depicted the helpless, paralysed feeling of the soldier as the shell approached him by sketching enormous weights chained to his feet. He could not move for fear. Emotion inhibited movement. In certain circumstances it must have meant safety to the animal to lie motionless, as still as the dead. Some species of animal react thus with comparative regularity. From this aspect at least, the phenomena of the affective cataplectic fit fall into line with reactions phylogenetically ancient, and are capable of psychological and teleological explanation.

But the difficulty remains: in clinical cases cataplexy results more commonly from activation on the side of laughter, and has indeed been observed when joy or gladness has provided the excitant. It is not easy to see how the hilarious mood should cause an immobility of biological advantage to the person concerned. Yet, once again, everyday parlance has the expression, ' I nearly died of laughing.' It would seem, perhaps, that the mechanism once utilized by fear to produce motionlessness can also be activated by an emotion of opposite quality, and that what has a biological meaning in one set of circumstances has little or none in another. The question, however, needs closer consideration than can be allotted to it in this place.

It is also worth remarking that the matter of unexpectedness has much to do with the onset of the cataplectic attack. If the subject is prepared beforehand, it may never evolve. Forewarned is forearmed in a physiological as well as in a military sense. The cortex can exert a corticifugal, cortico-thalamic inhibitory influence before thalamocortical impulses get their start. This fact of clinical observation has often been confirmed ; and it is of interest because it is not paralleled to anything like the same extent in the case of epilepsy. In the case of the latter, cortical inhibition counts almost for nothing. May we infer that corticothalamic inhibition has greater momentum than corticocortical inhibition ?

Cataplexy is a physiological experiment evolving before the clinician's eyes, and giving him a chance of insight into the working of neural mechanisms more revealing than that provided by epilepsy. He should take every opportunity of studying it minutely, for he has this great advantage over the experimentalist, that the phenomena of cataplexy are at present incapable of being reproduced in the laboratory.



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# PARESIS OF UPPER MOTOR NEURONE TYPE FOLLOWING HERPES ZOSTER

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PARALYSIS accompanying or following herpes zoster is comparatively infrequent although examples have from time to time been recorded. As a rule the paralyses are of the lower motor neurone type and may be classified according to their anatomical situation. Those most commonly met with are the oculo-motor, the facial, the brachial and the abdominal varieties. Crural paralysis is very rare, and only two cases appear so far to have been recorded. Hardy,[1] in 1876, reported a case in which atrophy of the calf muscles followed an attack of zoster. The eruption was situated over the course of the sciatic nerve. It extended throughout the whole length of the limb and lasted for 20 days, but the pain persisted longer. Four to six weeks from the beginning of the illness, there developed a complete amyotrophy of the muscles of the calf and of the external parts of the leg. Some degree of recovery eventually took place. Cornil[2] recorded the second case in a woman aged 58. Fifteen days after an attack of herpes zoster, which was confined to the thigh, atrophy of the muscles of the thigh was observed. The knee-jerk was completely abolished. Slight recovery of function took place but some atrophy and paresis persisted.

Cases of involvement of the upper motor neurone are rarely met with. Dumery (quoted by Baudouin and Lantuéjoul[3]), in 1896, noted a left hemiplegia following four weeks after a herpes zoster confined to the ophthalmic division of the right fifth nerve in a migrainous subject. Brissaud,[4] in 1890, referred to three cases of crossed hemiplegia in patients with ophthalmic or facial zoster. Bruce,[5] in 1907, described an unusual case in a woman aged 54. The rash was confined to the distribution of the seventh and eighth dorsal segments of the right side. Just over three weeks later, when the cutaneous lesions had healed, she noticed weakness and numbness of the right leg, and on examination she showed signs of involvement of the pyramidal tract on the right side together with an ipsolateral dissociated anæsthesia below the level of the rash. In addition, the patient had noticed an absence of sweating in this area. Three weeks later the left lower extremity was found to be involved in exactly the same way as the right. Gradual recovery followed and the patient was left with no disability other than troublesome paræsthesia in the lower limbs. Objectively there remained an undue briskness of the knee-jerks but no other abnormality. Karman,[6] in 1928, described a case

of unusual interest. The patient was a woman aged 59. For a year she had complained of pains in the lower lumbar region of the right side. She then developed the rash of herpes zoster over the distribution of the right sixth dorsal segment and this was followed shortly by paræsthesia in the right leg and astereognosis. Both legs became weak and by the end of the eighth week double incontinence had supervened. She complained of radicular pains accentuated by coughing and sneezing. There was tenderness on pressure over the fifth and sixth dorsal vertebræ and a spastic paraplegia with complete sensory loss, both exteroceptive and proprioceptive, below the level of the sixth dorsal segment. The cerebro-spinal fluid was reported to be normal in all respects and no block was demonstrable by the method of Quickenstedt. X-ray examination of the spine and spinal lipiodol were also negative. Laminectomy was performed over the third to fifth dorsal vertebræ and an intradural cyst was found at the level of the sixth dorsal segment (fourth dorsal vertebra). This was evacuated but the patient unfortunately died of septic meningitis and no post-mortem was available. Alajouanine and Bernard Griffith,[7] in 1931, described an interesting case in a female aged 26. At the age of 17 she had an eruption of herpes zoster on the right thigh in the distribution of the fourth lumbar segment. There was pain in this area but the illness was afebrile. Fifteen days later she experienced a temporary paresis of the right leg which improved considerably within eight days and had recovered completely at the end of three months. At the age of 21, she had a return of the pain in the right leg and of difficulty in walking. Two years later, at the age of 23, she exhibited typical signs of disseminated sclerosis.

## PERSONAL CASE

The patient, a male aged 58 years, developed herpes zoster in November, 1929. The eruption occurred without premonitory symptoms and involved the seventh thoracic segment on the left side; it was very profuse and extended almost continuously from the spine behind to the mid-line of the abdomen in front. The skin lesions followed the usual course but as they healed the patient complained of severe pain in the area involved, together with a sense of discomfort in that part of the abdomen lying below the herpetic zone. Three weeks later, cutaneous anæsthesia and analgesia were present over the area affected by the herpes—mainly the left seventh thoracic segment—and the pain continued with varying intensity. Six weeks after the onset of the rash he began to complain of severe pain in the left leg, together with inability to lift the limb. Muscular weakness of the left leg with increased deep reflexes was present but cutaneous sensation apparently normal; the right leg was unaffected and sphincter control was normal. X-ray examination of the spine showed no abnormality and the blood Wassermann reaction was negative. The pain in the herpetic area and in the leg both gradually disappeared, but the patient continued to complain of some hyperæsthesia of the left side of the abdomen in its upper part with a sense of numbness below. The weakness of the left leg slowly improved and at no time did the right leg show any abnormality. His condition when seen six months after the onset of the rash was as follows :—

A zone of cutaneous hyperæmia about 3 inches in width with slight patchy desquamation was still present over the site of the former herpes, extending from

within an inch of the spine behind to the midline in front, and corresponding to the total area supplied from the seventh thoracic segment.

Sensation : Cutaneous sensation over the above area was impaired to all stimuli. Elsewhere, including the skin of the abdomen below this level and over both legs, cutaneous sensation was normal. Vibration sense was impaired and shortened in duration over the patella, anterior surface of tibia and both malleoli in the left leg, but normal elsewhere.

All cranial nerves including the fundus oculi were normal.

There was distinct weakness on contraction of the abdominal muscles on the left side below the level of the seventh thoracic segment and particularly just below the herpetic area. The abdominal reflexes on the right side were brisk, but they were absent on the left side. The left leg was somewhat spastic and all muscular movements, though present, were diminished in power especially flexion at the hip and dorsiflexion at the ankle. There was no apparent wasting. The left knee and ankle jerks were greatly increased as compared with the right, which were rather sluggish but improved with reinforcement. Left ankle clonus of abortive type. Plantar stimulation gave a strong flexor response on the right side but an indefinite extensor reflex on the left, while Oppenheim's reflex was extensor on this side. Co-ordination in both legs was fairly good and Romberg's sign negative.

Other Systems : Heart and lungs normal ; palpable arteries in good condition ; pulse 76 regular ; blood-pressure 160/90. Liver and spleen impalpable ; urine no abnormality.

The cerebro-spinal fluid was clear, colourless and under normal pressure. The number of cells per c.mm. was 4 ; they were all small lymphocytes. Total protein 0·025 per cent. No excess of globulin. Lange and Wassermann reactions negative.

Two months later the patient was seen again and found to be walking much better. He still complained of a continual ache on the left side over the zone of the zoster and passing through to the back. All movements of the limbs were good including dorsiflexion of the hip and ankle, but there was a slight reduction in the power of all muscle groups of the limb. There was some impairment to light touch over the former herpetic area, which now showed scars. The deep reflexes of the left leg were still brisker than those of the right and the abdominal reflexes were diminished on the left. The left leg felt colder than the right but both dorsalis pedis and posterior tibial pulses were normal.

Six months later he was seen again (now 14 months after the onset of the rash). He still complained of aching on the left side and that his left leg would give out after walking for about a mile ; also that it would ache on exertion. The signs were almost as they were six months previously with the exception that the degree of spasticity was decidedly less. He has been seen again recently, now three and a half years after the onset of the rash. He remains much the same but is now able to walk further. An undue briskness of the deep reflexes of the left leg still persists.

## DISCUSSION

Within recent years there have appeared several valuable papers on the pathology of herpes zoster. In particular we would mention those of Lhermitte and Nicholas,[8 and 9] Fauré Beaulieu and Lhermitte,[10] Lhermitte and Vermès,[11] and Favre and Dechaume.[12] These authors emphasize the fact that the pathology is essentially a myelitis, the bulk of the changes being in the region of the posterior horn but that similar changes can occur either in the cord, medulla or brain. This is in contradistinction to the theory previously held, that of von Bärensprung [13] and of Head and Campbell,[14] that the essential lesion is an inflammation of the posterior root ganglion or

of their analogues—the cranial nerve ganglia. The modern view, therefore, is to regard herpes zoster as being primarily an acute posterior poliomyelitis, and further in support of this we have the occasional occurrence of a lymphocytosis early in the disease. In these circumstances, it is easy to see that the pyramidal tracts may occasionally be involved in the disease process.

In our case, the inflammation evidently spread from the posterior horn towards the lateral column and involved the motor fibres on their way downwards in the spinal segment concerned. The posterior columns also appear to have been affected to a slight extent as indicated by the impaired vibration sense, but no central grey matter was implicated as common sensation below the level of the lesion on both sides was normal.

In Bruce's case, the fact that the motor and sensory changes were on the same side of the body, indicates that the path for common sensation was involved before it had crossed to the opposite side of the cord. Moreover, the patient had noticed an absence of sweating in the parts affected, and Bruce attributed this to involvement of the intermediolateral tract. To account for these changes, Bruce suggested an inflammatory thrombosis of the postero-lateral or posterior cornual arteries, or perhaps the midcornual branch of the central artery. The subsequent extension to the other side in his case was accounted for by postulating the same changes on the other side of the cord.

As no case resembling either our own or Bruce's case has come to autopsy, the underlying pathology must needs be a matter for speculation. It is reasonable, however, to assume that the inflammatory process spreads from the posterior horn into the neighbouring structures. One would expect a considerable degree of recovery, as indeed occurred in both these cases, when the inflammatory process undergoes involution. Were the cause of the symptoms and signs thrombosis of the vessels, one would expect a lesser degree of recovery because the spinal arteries are end arteries.

Quite apart from giving rise to signs of spinal cord involvement, herpes zoster may also occur symptomatically as an expression of disease of the cord. Thus, in the case of Alajouanine and Bernard Griffith, to which reference has already been made, the herpes zoster apparently coincided, both in time and situation, with the first attack of a disseminated sclerosis. In Karman's [6] case, the herpes zoster coincided with the early symptoms of a spinal cord tumour. The other possibility in this interesting case, as Karman suggested, is that the intradural cyst was the result of the inflammatory process, i.e. secondary to the herpes zoster. There are also in the literature other interesting examples of herpes zoster coinciding with disease of the spinal cord. Schubach [15] has described herpes zoster in association with Landry's paralysis in a woman aged 62. Marinesco [16] also reported a zoster of the gluteal region with hyperacute myelitis of the conus medullaris in a man aged 36. The attack followed the seventh injection of a gonococcal vaccine which he was receiving for a urethritis acquired at the age of 22. It is of interest to note

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that this patient had had an acute anterior poliomyelitis at the age of five which had left him with a lower motor neurone paralysis of the left lower extremity. Becker [17] has described zoster coinciding with polyneuritis, the result of bismuth therapy in the treatment of syphilis. It would always seem advisable, therefore, in any case of herpes zoster to bear in mind the possibility that zoster may be symptomatic of some underlying disease of the central nervous system.

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# Short Notes and Clinical Cases

## COMPLETE DEGENERATION OF THE POSTERIOR COLUMNS OF THE SPINAL CORD WITH CHRONIC POLYNEURITIS IN A CASE OF WIDESPREAD CARCINOMATOUS DISEASE ELSEWHERE

By

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THE patient, a gardener, aged 39 years, was admitted to hospital on October 3, 1931, for pneumonia of the lower part of the left lung. There was no fever after October 10, and he was sent to a convalescent home on November 4. Whilst in the convalescent home he had paræsthesiæ (sensation of pricking) in the hands and feet, and on January 8, 1932, he was re-admitted to hospital for marked weakness of the lower extremities causing difficulty in walking; his knee-jerks were absent. The condition was supposed to be one of polyneuritis following pneumonia. His blood-serum gave negative Wassermann and Meinicke reactions. The blood-count showed nothing abnormal excepting that there was relative excess of lymphocytes in the differential count. The urine was free from albumin and sugar. By ophthalmoscopic examination the fundi appeared normal.

Under treatment by electricity, a little massage and liquor strychninæ the patient did not improve, and in the middle of February there was much muscular atrophy in his legs and hands. This muscular atrophy was decidedly more than could correspond to the wasting which was part of his general condition. The cerebrospinal fluid (February 24) gave positive Nonne-Apelt and Pandy reactions, negative Wassermann reaction and no pleocytosis. He complained of severe pains in his hands and feet, and there was much vomiting. At the beginning of March no tendon or periosteal reflexes could be obtained in the upper limbs. Sensations for pain and temperature were absent in both lower limbs up to above the knees but tactile sensation remained; the same sensory symptoms were noted in both hands. There was no disturbance of postural sense, but astereognosis was present in both hands.

Various drugs were tried without effect. At the end of April he complained greatly of burning sensations in the upper limbs distal to the elbows and in the whole of the lower limbs.

In May, owing to occasional vomiting, the gastric contents were examined

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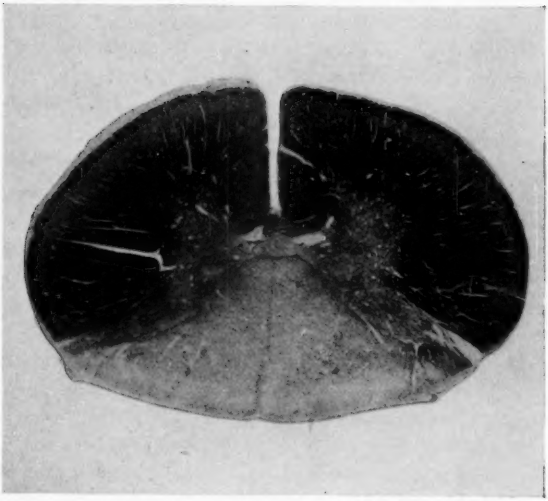
(*a*)

(*b*)



Transverse sections of (*a*) lower cervical region, and (*b*) mid-dorsal region of spinal cord, stained by Spielmeyer modification of Weigert's method, showing complete degeneration of posterior columns of Goll and Burdach.

but no achlorhydria nor anything abnormal was found. He could walk a little, but slowly; no 'high stepping' gait.

In September his general condition had much deteriorated and he could

eat hardly anything. A swelling of the left parotid salivary gland had gradually appeared. At the beginning of October the emaciation was great and there was still vomiting. On October 4 the pulse was 120 ; respiration 30 ; the temperature was 97·4° F., but there had been slight fever (100·1° F.) on the morning of the previous day. There was a leucocytosis of 19,000. He was being given occasional intravenous injections of a glucose solution. He died with great dyspnœa after one of these injections on October 6, 1932.

The *necropsy* showed extensive malignant neoplastic disease in both lungs (especially the left), the pancreas, both suprarenal glands (the right more enlarged than the left), the liver (especially the left lobe), both kidneys, the spleen (which was moderately enlarged, weighing 250 grammes), and the upper part of the vertebral column ; the enlarged left parotid salivary gland was likewise the site of neoplastic infiltration. Some of the abdominal lymphatic glands were infiltrated and there were a few minute metastases in the peritoneum over the colon.

There was no involvement anywhere of the spinal meninges. The whole of the spinal cord was removed for further examination.

Microscopic examination of the tumours showed that they were all of the same histological structure, resembling the so-called oat-celled carcinoma of the lung.

Unfortunately the peripheral nervous system was not examined, but transverse microscopical sections of the spinal cord were made from : (1) the lower cervical region ; (2) the mid dorsal region ; and (3) the lumbar enlargement. Sections were stained by the Spielmeyer modification of the Weigert method and by hæmatoxylin and eosin. The sections of the cord showed complete—or practically complete—degeneration of the posterior columns in all regions, cervical to lumbar. The grey matter of the cord and the other ascending and descending tracts were normal. There was no sign of any gross inflammatory reaction in any section, and no engorgement or ' cuffing ' of vessels, though macrogliosis was evident in the degenerate area. The pathological picture suggested a degeneration of the posterior columns, secondary to some general cause, which had produced widespread demyelination and disappearance of axis-cylinders ; the degeneration was equally severe in all sections.

## REMARKS

The vomiting may have been largely due to suprarenal involvement, but one can hardly say how long the carcinomatosis had been in progress. Although there was no post-mortem examination of the peripheral nervous system, it is fairly certain that genuine polyneuritis was present as well as the practically complete degeneration of the posterior columns of the spinal cord, for there was nothing in the spinal cord that could have caused the severe muscular atrophy in the legs and hands. It is very likely that the carcinomatous disease had already commenced at the time of the onset of the first

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nervous symptoms (viz., the paræsthesiæ in the hands and feet) soon after the patient's recovery from the pneumonia at the end of 1931. The pneumonia itself may have been on a local carcinomatous basis. One is inclined to think that the degeneration of the posterior columns of the spinal cord was secondary to a carcinomatous toxæmia, but there seems to be no great support for such an assumption in the literature of the subject, though it is acknowledged that polyneuritis may develop on the basis of a carcinomatous cachexia (compare H. Oppenheim, *Nervenkrankheiten*, 7th German edition, Berlin, 1923, p. 821). It is, however, probably not uncommon for varying degrees of involvement of the spinal cord to occur in various forms of polyneuritis, for example, in lead and diphtheritic neuritis.

We have to thank Professor J. C. G. Ledingham for having so kindly provided us with microphotographs illustrating the sections of the spinal cord.

# Editorial

## MICRENCEPHALY

THERE is a curious tendency of the human mind to be attracted by things which are rare or bizarre, while others—albeit the more important—if commonplace and conforming to a general type, completely fail to excite interest and are fated to slip into the limbo of dull and forgotten experience. Even such a humble claim to oddness as size excites the attention towards an object, should it be larger or smaller than its average fellows. A practical example of this tendency is afforded by the establishment of a special type of amentia based on the arbitrary selection of one particular anatomical feature, namely, the smallness of the head. For this the term microcephaly has been introduced, though as the small-headed person must of necessity possess a small brain, it would be preferable to employ the designation ' micrencephaly.' Exactly what constitutes a small head has never been definitely settled. Ireland suggested that all crania with a maximum circumference of 17 inches or less should be included, but such extreme examples are seldom seen ; and it is now more or less generally agreed that a measurement of 2 or 3 inches more does not necessarily exclude the subject from this category, some authorities even claiming that size is of less importance than shape. Here, however, it must be confessed that apart from the small size of the head there is clinically little which is distinctive. A small brain may, of course, be an instrument of exceptional human success, for the tissues of the human organism must be interpreted in terms of quality as well as quantity. Ethnology teaches us that some of the small-headed races have yet been the most successful in the development of the fine arts of the goldsmith, the engraver and the potter. In their construction of tombs and temples and their knowledge of ornamental arts, the old Peruvians were from that standpoint not far behind the civilization of to-day. Yet they possessed a brain no larger than that of the Hottentot, and far below that of the barbarous hordes of their own race. We cannot affirm, therefore, that

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the races which have shown most intellectual force have had the largest heads, but on the whole the prejudice in favour of heavy-brained persons is not ill-founded.

Although from the earliest times microcephalics have been the objects of ridicule or curiosity, it was not until the middle of last century that the peculiarities of this interesting type became the object of serious study. At this period, physicians with an enthusiasm which did more credit to their imagination than to their clinical acumen were largely concerned with the description of special clinical types of idiocy, among which the microcephalic from his fancied pithecoid resemblance came to have pride of place. At a time when Charcot was holding his audiences in the Salpêtrière enthralled by the theatrical display of manifestations of *la grande hystèrie*, Lombroso was achieving equal notoriety in Italy with his small-headed idiots—l'uomo uccello, the bird-man who chirped and used his arms like wings; l'uomo coniglio, the rabbit-man who stamped with his feet as rabbits do; il scimmin, the monkey-man who leapt in the air like an ape, and others of the like, became objects of popular interest and formed the subjects of numerous mémoires, besides opening up a fertile field for theorists, who, biassed by the study of particular instances, were not loath to avail themselves of the opportunity of propounding widely different hypotheses. At least three of these deserve notice.

The occasional occurrence among microcephalic idiots of hypertrophied scalps, which seemed too large for the skulls they covered, and the report by Baillarger of early closure of the cranial sutures, suggested that a small brain was to be accounted for by premature synostosis of the bones of the skull. This could be remedied by heroic operative procedures, such as the removal of long strips of bone along the cranial sutures. Useless in all other directions, these, at least, served to dispel confidence in what proved to be an unwarrantable assumption. Another view, popular at that time, and one with which the name of Virchow is particularly associated, held that idiots of this type represented the return to the simian stage. Acceptance of this view is rendered difficult by the unsatisfactory character of the anatomical findings. Far from presenting constant features, microcephalic brains neither resemble one another, nor the brains of any monkey, save in those common features in which the simian brain resembles the normal human one.

A third theory assumed that microcephaly is explicable

as a simple curtailment of development, and for this view there is a good deal to be said. The mentally abnormal tend to have abnormal heads, and just as there are giants and dwarfs as well as individuals of normal stature, so too, we may expect to meet the extremes in head size—the microcephalic and the macrocephalic.

Considered from this standpoint, microcephaly merely represents the extreme degree of small-headedness, a suggestion which becomes the more plausible when we recognize with Professor Berry that some degree of small-headedness is present in two-thirds of all mental defectives. Be this as it may, it has, of course, long been known to embryologists that the susceptibility of a given part to noxious agents varies directly with the general physiological activity of the protoplasm. In the human embryo it is the head which is the region of greatest activity, and which is most susceptible to inhibiting agents. As the researches of Donaldson and others have shown, cell division in the nervous system commences as early as the end of the second week of embryonic life, and theoretically there is no difficulty in supposing that the process of normal cerebral development may simply stop when a certain stage has been reached. It is thus possible, as Child contends, to regard microcephaly as a characteristic result of differential inhibition along the polar axis, but until recently we have been entirely in the dark as to what the nature of these inhibiting influences might be. Some recent reports, however, suggest that there is now available convincing evidence that the development of the embryonic or fœtal nervous system may be arrested by external agents.

In 1920 Aschenheim described a four-year-old child with microcephaly, intellectual retardation and eye malformation, whose mother had been exposed to X-ray irradiation during pregnancy. Since then a number of foreign observers have placed additional cases on record, which brings the total number of cases of ' radiogenic microcephaly ' up to rather more than 20. Murphy and Goldstein, whose researches form a valuable contribution to this subject, found in a series of 75 children irradiated during fœtal life that approximately one-quarter were microcephalic, and they conclude that irradiation must act somewhat similarly to toxic factors or environmental influences in arresting fœtal development. This is, perhaps, a somewhat large assumption, since the X-rays and the emanation from radium represent a very special form of physical energy, which so far as we know is not likely to affect the

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human subject injuriously unless specially directed and controlled to that end. Yet it must not be forgotten that in lower animals the various modifications of development which can be produced experimentally are non-specific as regards agents: they can all be produced by anything which will inhibit general protoplasmic activity. In fishes, for example, microcephaly can be readily brought about by slight chemical alterations of the fluid medium in which they are developing. Furthermore, in the experiments of Foersterling on rabbits, X-ray irradiation produced ocular disturbances, a retarded growth of the skull and an altered shape of the nose, results which suggest for all practical purposes that the pathological changes in the experimentally irradiated animal are analogous to the disturbances observed in the child irradiated in utero. In the present state of our knowledge, facts such as the foregoing, although suggestive, cannot give conclusive evidence of the environmental origin of microcephaly. They do, however, seem to indicate the extreme susceptibility of the nervous system to external agents, and it is at least permissible to claim that the studies of Murphy and Goldstein have given an impetus to the further study of this interesting condition. It may be anticipated, indeed, that ere long these will yield results which will be of more value than the tangled web of theory spun by previous workers in this field.

# Abstracts

## Neurology

### NEURO-ANATOMY AND NEUROPHYSIOLOGY

[1] **The reflex of posture and shortening** (Incertezza e difficolta nel rilievo dei reflessi di postura e di accorciamento).—S. GARETTO. *Riv. di pat. nerv. e ment.*, 1932, **39**, 559.

THE author has studied the postural reflex of shortening of the foot and of the thigh in normal subjects and in patients suffering from various types of nervous and mental illness, and points out the difficulty which is met with in the interpretation of such phenomena. Basing his results on his observations he concludes that the posture reflex is not constant in the normal subject, that in pyramidal lesions it is not always abolished or diminished, and that in extrapyramidal lesions the reflex hypertonia of posture is not always found.

With reference to the postural reflex of the catatonic type in subjects who are not catatonic he says that such phenomena may be attributed in the Parkinsonian syndrome to an exaggerated postural reflex allied in great measure to hypertonia, while in the syndromes where hypertonia is absent it may be regarded as a psychomotor reaction, which is not exactly the same as a catatonic syndrome. As to the sign of anticipation of passive movement, the author denies to such a phenomenon any special morbid significance and says that it has nothing in common with postural reflexes.

R. G. G.

### NEUROPATHOLOGY

[2] **Fatty substances in the neurinomas of the acoustic nerve** (Sulle sostanze grasse dei neurinomi del nervo acustico; richerche istopatologiche ed istochimiche).—U. DE GIACOMO. *Schweiz. Atch. f. Neurol. u. Psychiat.*, 1933, **31**, 73.

IT is extremely common to find fatty material in eighth nerve tumours, either lying free in the syncytial plasma of the growth or with a certain predilection for a perinuclear site. Occasionally it fills the whole cytoplasm. The cells in which this occurs are termed by the author 'xanthomatoid'; they are generally situated near vessels but also under the capsule of the



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neoplasm. Chemically, the fat consists not of stearine but of phosphatides. Hence 'lipoid metamorphosis' is a more accurate expression than 'fatty degeneration.'

S. A. K. W.

[3] **Neurolysis in the cerebrospinal fluid** (Il processo neurolitico nel liquido cerebrospinale umano).—O. Meco. *Rassegna di studi psichiat.*, 1933, **22**, 214.

In human spinal fluid is present the property of disintegrating neural tissue, a neurolytic potentiality which was found in 78 per cent. of 246 fluids examined by the author. This property is not destroyed by inactivation, filtration, boiling, or centrifugalizing. It is principally found in relation to pathological conditions of the cortex rather than of white matter. The phenomenon is probably due to the presence of unknown products of cellular activity. If the products of neural disintegration have been shed into the fluid the neurolytic power of the latter is increased.

J. V.

[4] **Considerations on the pathogenesis and diagnostic value of eosinophilia in the cerebrospinal fluid** (Considerazioni sulla patogenesi e sul valore diagnostico della eosinofilia del liquido cefalo-rachidiano).—A. Mari. *Riv. di pat. nerv e ment.*, 1932, **39**, 317.

The author considers that the presence of eosinophils in the cerebrospinal fluid is due to a disturbance in the gaseous exchange of the tissues with which the fluid comes in contact. The only condition in which it is found is in cysticercus of the central nervous system, hence the presence of cells is of great diagnostic importance.

R. G. G.

[5] **Lange's reaction in neurosyphilis** (La reaxione di Lange nella neurosifilide).—L. Cabitto. *Riv. di pat. nerv. e ment.*, 1932, **39**, 145.

The author compared Lange's reaction and the benzoin reaction and considers that the former is very sensitive in the diagnosis of paresis and neurosyphilis and superior to the reactions of the colloidal resinous bases. He thinks that it is still more sensitive if prepared with a 2 per cent. chloride of gold instead of the 1 per cent. proposed by Lange.

R. G. G.

[6] **The permeability of the blood-cerebrospinal-fluid filter** (Sulla permeabilita del filtro neuro-ematico).—P. Ottonello. *Riv. di pat. nerv. e ment.*, 1932, **39**, 592.

Following a technique which seems to exclude error the author has shown in dogs that the inoculation of *Trypanosoma Brucei* in the spinal subarachnoid

space confirms the permeability of the blood and cerebrospinal-fluid filter for protozoa, already demonstrated for the plasmodium vivax.

R. G. G.

[7] **Histopathological characteristics of peripheral nerves in amputated extremities of patients with arteriosclerosis.**—J. B. PRIESTLEY. *Jour. Nerv. Ment. Dis.*, 1932, **75**, 137.

IN the peripheral nerves of extremities amputated for arteriosclerotic gangrene are evidences of definite Wallerian degeneration and fibrosis. These changes appear more pronounced in the distal than in the proximal portions of a given nerve, and the degree of change is directly proportional to the degree of arteriosclerotic alteration of the vessels. This suggests a peripheral origin for the degeneration, which probably depends on ischæmia for its production. In support of this supposition is the fact that Brown, Allen, and Mahorner, studying thromboangiitis obliterans, found that somewhat similar changes in nerves existed with occlusive disease of the vessels. Woltman and Wilder, studying nerves in cases of diabetes mellitus, attributed the change in nerves to arteriosclerosis of the vessels present. Ischæmia of nerve fibres from numerous causes may result in pain of varying intensity, from mild neuritis to the excruciating pain typical of sudden vascular occlusion, and the ischæmia may represent the etiological factor in many cases of neuritis. To support this view one has only to observe the clinical and symptomatic improvement resulting from treatment directed toward increasing the efficiency of the local circulation in these conditions.

R. G. G.

[8] **The value of the oscillation of acid base equilibrium for determining the occurrence of epileptic convulsions** (Il valore delle oscillazioni nell'equilibrio acido-basico per lo scoppio dell'accesso convulsivo epilettico).—C. POLI. *Riv. di pat. nerv. e ment.*, 1932, **39**, 100.

WHILE it is established that oscillations in the acid-base equilibrium are attendant on convulsive seizures, it is not yet certain whether these are causal or merely a biochemical change accompanying the fit.

R. G. G.

## SENSORIMOTOR NEUROLOGY

[9] **A clinico-pathological study of the intracranial arachnoid membrane.**—L. DAVIS and H. HAVEN. *Jour. Nerv. Ment. Dis.*, 1931, **73**, 129.

A SERIES of 10 cases showing definite histopathological changes in the arachnoid membrane are presented. These include one verified intracranial



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tumour, two with the pathological process involving the optic chiasma, and seven with the clinical picture of an intracranial neoplasm in which no tumour was verified at operation. Each of these showed definite changes in the arachnoid membrane, which can be classified into three different groups according to the histopathological picture each presented. Two of the cases gave a history of trauma sufficient to be considered as an etiological factor. No attempt is made to discuss the etiology of the other cases.

The authors believe it is better to classify these patients as 'tumour suspects' than to make a diagnosis of arachnoiditis. In this manner they are not apt to be dismissed from future careful and close observation for the early evidence of symptoms of more serious intracranial pathology. At present these clinical and surgical findings should not be classified as an entity, although the continuous observation and post-mortem verification of these lesions multiplied many times may lead to such a conclusion.

For the purpose of classifying the histopathology encountered under such circumstances in the future they suggest three main types: inflammatory, fibrotic and hyperplastic.

R. G. G.

[10] **Toxic degenerative myeloradiculitis due to large round-celled carcinoma of the ovaries** (Mielo-radicolite tossico degenerativa da carcinoma a grandi cellule rotonde dell'ovaio).—V. TRONCONI. *Riv. di pat. nerv. e ment.*, 1930, **36**, 304.

A CASE of atypical neoplasm is discussed, a large round-celled carcinoma of the ovary with a rapid course and diffuse lymphatic metastases. The morbid picture of the neoplasm was complicated by the appearance of a subacute condition involving the spinal medulla in the lumbosacral region and the corresponding roots. Pathologically, the spinal lesion was more evident than the root lesion and had the appearance of a degenerative toxic myeloradiculitis. All other causes of this condition, including that of grave anæmia, were excluded; presumably the toxic factor was derived from the neoplasm, but how this acted is uncertain.

R. G. G.

[11] **Cortical herniations.**—C. T. PROUT. *Jour. Nerv. Ment. Dis.*, 1931, **74**, 468.

MULTIPLE herniations of the brain are caused by increased intracranial pressure, but they are not always present in such a condition. Their presence is dependent not only on the duration of the increased intracranial pressure, but on its severity and the rapidity of its onset. They are most commonly found in the presence of tumour of the brain. Age is not an important factor, except that in advanced age there is a greater tendency to increased blood

pressure, and to temporary episodes of increased intracranial pressure. They are of importance from the standpoint of X-ray diagnosis, because they must be distinguished from malignant tumours which erode the inner plate of the skull. Their situation is not important in localizing lines of pressure. Evidence points to the fact that they follow the villi, although they do not necessarily follow them in the presence of other dural weakness.

R. G. G.

[12] **On the problem of traumatic parkinsonism.**—A. E. KULKOV. *Jour. Nerv. Ment. Dis.*, 1932, **75**, 361.

THIS is a discussion of the possibility of trauma as a cause of Parkinsonism. Two cases are quoted in which there would seem to be a clear and continuous history from the trauma to the full development of the syndrome, but in considering the whole question the following points must be taken into account.

The trauma must be severe enough to be able to cause some cranial lesion or at least call forth symptoms of a commotio cerebri.

The patient who suffered from the trauma must not have been previously subject to any cerebral symptoms.

The Parkinsonian symptoms must not follow the trauma immediately; their development ought to be preceded by some prodromal symptoms (cerebral symptoms) which would then gradually develop a characteristic clinical picture. Bing believes that, should these points be strictly observed, the number of cases of traumatic Parkinsonism would be reduced to a minimum.

But the author thinks that if he could add a clear and precise picture of an objective previous history showing the course of the disease from the very moment of the accident (trauma) to the onset of Parkinsonism, tracing it point by point, and considering it chiefly from the standpoint of the possibility of a former occurrence of a rudimentary form of encephalitis (in connexion with the trauma), the number of cases of 'pure,' unquestionable traumatic Parkinsonism would be still more reduced.

R. G. G.

[13] **Cysticercosis as a cause of epilepsy in man.**—W. P. MACARTHUR. *Trans. Roy. Soc. Trop. Med.*, 1933, **26**, 525.

THIS is a short communication with the object of redirecting attention to human cysticercosis as a frequent cause of epilepsy.

The writer refers to the number of men finally invalided from the army on account of epilepsy and to the relatively large proportion who developed this disease in adult life during or after service abroad. With a view to

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determine to what extent cysticercosis is responsible for late-developing epilepsy in soldiers, all cases of epilepsy were submitted to a special investigation. Of 22 cases of epilepsy admitted to Millbank, 10 have been proved to be suffering from cysticercosis, while the history in a number of others was suggestive of parasitization. The author states that many cases have been missed in the past; he has himself encountered over 20 proved cases and has received reports of a number of others.

The process of invasion gives rise to no general reaction, the subject merely noticing the gradual development of small subcutaneous or intramuscular swellings. An ascertainable history of infestation with a tapeworm is rare; sometimes the fits commence at about the time the cysts are first detected; in other cases there may be a quiescent period between the appearance of the cysts and the first seizure; or cysts may be palpable only subsequent to the onset of fits. The seizures may be Jacksonian in type or resemble those of major epilepsy. The most helpful sign in diagnosis is the development of palpable cysts in the subcutaneous tissues or muscles. They would appear to be common in any part of the body. The completely calcified cyst gives a characteristic appearance on X-ray examination. The large number and wide distribution of cysts in three brains examined post mortem would not favour a resort to surgical measures.

The author adds that the importance of cysticercosis as a cause of epilepsy is not sufficiently realized either in the service or by practitioners in the tropics. The onset of epileptic seizures in a previously normal adult during or after service or residence abroad should suggest the possibility of this condition.

W. A. T.

## PROGNOSIS AND TREATMENT

[14] **The borate treatment of epilepsy** (Die Behandlung der Epilepsie mit Borsalzen).—Th. Bovet. *Schweiz. Arch. f. Neurol. u. Psychiat.*, 1933, **31**, 25.

A series of 72 severe epileptic cases of all kinds were treated by the borates. Of these, 11 benefited greatly, 13 moderately, and in three the effect was transient. Sodium borotartrate does best, and the corresponding potassium salt next best. Boric preparations can be given in combination with luminal or bromide. They seem to produce their best effect in patients of asthenic or schizoid type.

This brief article is interesting also from its historical references to the use of borax in epilepsy, long antedating its introduction by Gowers in England.

S. A. K. W.

# Psychopathology

## PSYCHOLOGY

[15] **Localization of consciousness and emotion.**—LELAND B. ALFORD. *Amer. Jour. Psychiat.*, 1933, **12**, 789.

A SERIES of cases exhibiting lesions in different parts of the brain were studied for any changes caused in consciousness and emotion or what seemed to be the physiological counterparts of these faculties. In no lesion of the right hemisphere (in right-handed persons) was any consistent disturbance in these two faculties observed. Not one of 33 cases of left hemiplegia exhibited confusion and only three an equivocal emotional instability. Lesions of the frontal, parietal, occipital and temporal lobes on the left similarly caused no definite disorder of consciousness and emotion. In an extensive vascular lesion of Head's speech zone, despite jargonaphasia and apraxia, the patient remained generally well-balanced, alert to events about him, and capable of appreciating their general import. Hence the conclusion that emotion and consciousness (in the sense of awareness alone) were not affected and therefore do not reside in this part of the cortex. It was eminently in the right hemiplegias, which generally arise from left capsular injuries, that definite and permanent confusion of consciousness was found (27 out of 55 cases of old hemiplegia). It seems, therefore, that an area somewhere in the left base is concerned with the maintenance of awareness. In these same right hemiplegias and in other lesions that certainly or inferentially affect basal structures directly or indirectly were found restlessness, irritability, delirium and other forms of instability, thus indicating that emotion is localized in the base and possibly is confined to the left side. All the conclusions are regarded as tentative and subject to the light of further evidence. The method of following psychological faculties through a series of brain injuries is believed to be a useful one and worthy of wider employment.

C. S. R.

[16] **The social psychology of time.**—NATHAN ISRAELI. *Jour. Abnorm. Soc. Psychol.*, 1932, **27**, 209.

IN one experiment intellectual judgements were made by 607 subjects on the importance of the different time divisions, all of which were compared with one another. By simply totalling the percentages of those who favoured each time division, it was determined that the combined groups regard the present as 1·2 times as important as the future, and 12·7 times as important as the past. In a second experiment it was found that the past is hardly important in the emotional time behaviour of the subjects; while only 13 per cent. worry much about their own past misfortunes, 94·5 per cent. are

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hopeful about their own future; 67·4 per cent. worry about present problems, 56·3 per cent. daydream about things that are going to happen, and 89·3 per cent. often shape their plans in advance. Thus it has been shown that the future is of most and the past of least importance in the orientation of the subjects examined.

C. S. R.

**[17] Erethizophrenia and kolyphrenia* : a physiological conception of psychological types and their relation to psychopathology.**—J. RAMSAY HUNT. *Amer. Jour. Psychiat.*, 1932, **12**, 475.

THE author formulates the conception that states of exaggerated cortical excitability (erethizophrenia) and cortical inhibitability (kolyphrenia) are the important factors in the manifestations of psychological type and many of the disorders of mental function which are encountered in abnormal psychology.

Experimental investigations have shown that the whole vegetative mechanism is supplied by inhibitory and excitatory fibres which cooperate in vegetative function. No such inhibitory nerves are recognized in the central nervous system. Here it is assumed that the nerve-impulse is blocked at the synaptic membrane of the excitatory cell, and that there are no specific inhibitory cells.

In 1923, the author proposed a *specific cell theory* of inhibition, based on pathological investigations of the corpus striatum. These studies showed that destruction of the small cells of the neostriatum was associated with a loss of inhibition within this ganglion, in the form of chorea. As these small cells of Golgi are widely distributed throughout the central nervous system and exist in every aggregation of grey matter, including the granular layers of the cerebral cortex, the specific cell theory was postulated, thus bringing the mechanism of inhibition in the central nervous system into harmony with that of the vegetative system, where both excitatory and inhibitory neurones exist.

According to Pavlov, the cerebral cortex represents a huge functional mosaic made up of shifting points of excitation and inhibition, closely intermingled. He also found a wide variation in the nature of the individual nervous system in his experimental animals and that two definite types, an excitable and an inhibitable, stand out with special prominence.

*Types of Psychic Excitability and Inhibitability in Man.*—In 1928, the author described two fundamental psychological types in man, an excitable or erethitic and an inhibitable or kolytic, based on the predominance of one or other of these opposing functions in the psychic sphere.

The *erethitic* type is characterized by a great activity of mind and body. Such individuals are very responsive, impulsive and easily excited. They

* Derived from the Greek: *Erethizo*, to excite, to provoke, and *phren*, the mind. *Kolyo*, to prevent, to check, and *phren*, the mind.

are quick-tempered, highly emotional and inclined to be restless. The kolytic type, on the other hand, is more calm and controlled, with a greater passivity of mind and body. They are slow-tempered and reflective, with a marked tendency to subjectivity and day-dreaming. The general characteristics which distinguish these types may also be recognized in the classifications of Jordan, Jung and Kretschmer.

The author regards it as significant that in all of these studies, although approached from very different angles, two fundamental and contrasting types are recognized, all very similar in their general outline and essential characteristics.

The following points are emphasized :—

1. The cerebral cortex which is concerned with psychic function is subject to the same physiological laws of excitation and inhibition as are the lower centres.

2. While the nature and localization of inhibition are still disputed questions there is neuropathological evidence supporting the theory of specific excitatory and inhibitory neurones in the central nervous system harmonizing with the existence of such a dual mechanism in the vegetative system.

3. Two fundamental psychological types may be recognized—an *erethitic* and a *kolytic*—due to a predominance of one or the other of these functions in the psychic field.

4. Exaggerated states of cortical excitation and inhibition, general or local, constant or transitory, are responsible for many of the well-known disorders of psychopathology.

AUTHOR'S ABSTRACT.

[18] **Measuring social adequacy.**—MARY JOSEPHINE MCCORMICK. *Mental Hygiene*, 1933, **17**, 100.

SOCIAL adequacy is defined as ' the quality by which a family is able to preserve its domestic life without unusual aid from the community.' The object of this study was to devise a measuring instrument adapted to the quantitative study of social adequacy. After a review of previous studies of home and environmental conditions, the rating-scale method was rejected in favour of the personal interview. The technique finally adopted involved a form to be filled out by a social worker after a personal visit to the home. In standardizing this scale, 249 casts were used, distributed among 14 groups chosen to represent widely different degrees of social adequacy. The 14 groups were ranked in accordance with their characteristic degree of social adequacy by 25 social workers and teachers of social science. The groups so rated were used as a criterion against which to validate separate items, and the scale as a whole. In the final scale, all items were omitted that did not yield coefficients either greater than + 0·30 or less than — 0·30. After omitting

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the families in which the data were seriously incomplete, a group of 184 families were left. These were all scored in this scale. The reliability was found by the method of split halves to be 0·955. The validity was 0·928. On the basis of the scores received by the different groups studied, it was possible to set up tentative norms corresponding to the various scores possible on the scale.

C. S. R.

[19] **A note on the effect of repeated hypnotic stimulation.**—O. H. Mowrer. *Jour. Abnorm. Soc. Psychol.*, 1932, **27**, 60.

Experiments were made on a rooster that had proved to be a valuable hypnotic subject. In view of the dearth of data on the practice effect of hypnosis, it seemed worth while to try to ascertain whether the duration of the hypnotic trance decreases or increases with repetition. The time required to induce the trance was counted from the onset of stimulation until the eyes had remained closed for 30 seconds. The duration of the trance itself was counted from the termination of stimulation until the bird got to his feet, which ordinarily occurred as soon as he opened his eyes. The intervals between the successive trials on a given day usually amounted to less than a minute, sometimes only to a few seconds. Although there was no significant change in the time required to produce a 30-second closure of the eyes, the duration of the trance dropped very markedly, in the end virtually to zero. This decrease appears to agree in general with certain observations which have been made on human subjects, and seems to offer another example wherein repetition, instead of 'fixating' or 'stamping in' a response in accordance with the traditional laws of learning, actually abolishes it. If it can be definitely established by further experimentation that the duration of the hypnotic trance customarily decreases with repeated stimulation, it may afford one of the many missing links in the explanation of hypnotic phenomena in general; certainly it will demand a reformulation of the so-called law of repetition.

C. S. R.

[20] **Association in normal and subnormal adolescents.**—Edna Willis McElwee. *Amer. Jour. Psychiat.*, 1931, **11**, 311.

Normal adolescents had a higher number of individual reactions than children under 16 years of age taken as a group. This fact suggests the presence of some emotional instability during adolescence. The subnormal adolescents had a much greater number of individual reactions than the normal adolescents. This may be explained in two ways: (*a*) The subnormal adolescents are more emotionally unstable during adolescence. (*b*) The subnormal adolescents should not be measured by the standard for children under 16, but for children under 10. On the latter standard a larger number

of individual reactions is permissible and greater abnormality in the quality of the reactions. With this interpretation the emotional instability of the two groups would be more comparable.

C. S. R.

[21] **An experimental study in hypnotic anæsthesia.**—JOHN B. DYNES. *Jour. Abnorm. Soc. Psychol.*, 1932, **27**, 79.

The particular point which this research shows quite clearly is the fact that bodily mechanisms as represented by respiratory and cardiac activity show little or no disturbance in their normal rate and rhythm following sensory stimuli in the hypnotic trance. In the normal waking state these same indicators invariably showed a disturbance in both rate and rhythm. The rate was accelerated and the subjects showed gross bodily movements as reflex responses to stimuli. The psychogalvanic reflex was little influenced by the hypnotic trance. A slight decrease, as compared with the normal, was noted in the trance following sensory stimuli. The verbal report of the subject as to the sensations perceived was substantiated by the objective record in both the waking and trance states. The hypnotic trance acts as a quieting influence and the usual response to strong sensory stimuli is inhibited.

C. S. R.

## NEUROSES AND PSYCHONEUROSES

[22] **Stuttering and allied speech defects.**—BARUCH SILVERMAN. *Canadian Med. Assoc. Jour.*, 1932, **27**, 268.

PHYSICAL defects of the mouth, teeth, tongue, nose and throat should be corrected and the principles of physical hygiene given due consideration. The child should be encouraged to develop the habit of using words correctly, to name objects, and especially to ask for things he wants, rather than resort to baby talk or have his needs so anticipated that he has no occasion for the use of words. He should be discouraged from imitating the stuttering speech of another child or adult, since occasionally such imitation leads to the development of a serious speech defect. Left-handed children should be allowed to use their hand of preference in the performance of any task, especially writing; and a child who is predominantly left-handed should not be urged to use his right hand, as such a procedure may interfere with normal speech development and not infrequently proves to be a cause of stuttering. Since stuttering is so often an indication of a person's inability to meet the requirements of the group in which he lives, it is most important that the social environment in which the child lives should be as free as possible of unnecessary excitement, emotional outbursts, fears, jealousies,

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persistent nagging or undue harshness, for such an environment tends to undermine a child's feeling of security and self-confidence, and interferes with his tendency to normal self-expression through speech.

C. S. R.

[23] **Functional disturbance of psychogenic nature.**—FRANZ ALEXANDER. *Jour. Amer. Med. Assoc.*, 1933, **100,** 469.

AFTER a discussion of the influence of psychic factors in physiological changes the psychogenic disturbance of the organs is defined precisely. Functional disturbances, though essentially identical with psychophysiological processes of expressing emotions such as laughter and weeping, are nevertheless distinct from them by three characters: (1) In the case of a psychogenic disturbance of an organ the emotion which seeks expression is unconscious, i.e., repressed. (2) The psychogenic disturbance is an unusual or an incomplete expression of a psychic tension. (3) The symptom is not able to relieve the psychic tension in the same way as normal psychomotor reflexes do. The repressed tendency, excluded from consciousness and thus from normal expression, sustains a steady tension which is the cause of the chronic dysfunction. Every psychic tendency seeks an adequate bodily expression. Normally this is through the conscious ego, probably localized anatomically and physiologically in the cortex. If this passage is blocked an unusual relief comparable to a short circuit takes place; this is the hysterical dysfunction of the organs. The investigation of the psychic constellation of the patient is important even in cases of certain organic disturbances since chronic functional disturbances may lead to real organic disorder.

For illustration the etiological problem of peptic ulcer is reviewed. In several cases the author has established the psychic origin of functional stomach troubles that preceded clinically diagnosed peptic ulcers and in one analyzed case it was possible to reconstruct the psychological factors that had been in the background of the stomach symptoms for many years. The repressed wish for a revival of the infantile relation of the child to the mother, maintains a permanent tension which tends to influence stomach functions. The associated wish to be fed, independent of hunger, serves as an unconscious substitute for the wish to be loved. Hence the stomach is exposed to a chronic and specific stimulus. In this way a chronic hypersecretion or hypermotility may arise. Alexander considers that it is established that many organic cases develop because there is an originating chronic psychic stimulus in the form of a repressed unconscious psychic tendency; that this leads to functional disturbance of an organ; and that a last phase manifests itself in organic morphological changes. The choice of the organ that serves for the expression of unconscious tendencies depends on the specific nature of the tendency. Just as passive receptive claims to be loved have an intimate

relation to the alimentary tract, so do fear and anxiety seem to have a close connexion with the circulatory system, whereas the emotions of spite, stubbornness or rejection of the environment may frequently, if they are repressed, find expression in the disturbance of the sphincter-function of the anus. Psychotherapy has mainly a prophylactic value. It consists in giving the patient the possibility of relieving psychic tendencies in the normal way by making conscious those repressed tendencies which, on account of repression, could not be expressed normally and had to find unusual pathological expression.

C. S. R.

## PSYCHOSES

[24] **Children of schizophrenic parents.**—EDNA M. LAMPSON. *Mental Hygiene*, 1933, **17**, 82.

THE significant questions arising from the data obtained from the study of approximately 200 families are: to what extent is inherited instability involved in the lives of the offspring, and in what manner has it manifested itself? The first fact to be noted is that 30 per cent. of the offspring have encountered difficulties of both a mental and a social nature. As these difficulties were diversified, they are designated as 'maladjustments' by the writer. Thirty-eight, or 50 per cent. of the psychotic parents, have had maladjusted children. Fifty-four per cent. of all the children in these 38 families were maladjusted. Varying factors involved in the disrupted homes have to be taken into account, as well as the so-called 'bad' homes. It was not clear that these two conditions alone were accountable for the innate or early acquired forms of maladjustment observed. It would seem that the instability resulting from the schizophrenic parent is a factor to be considered.

C. S. R.

[25] **Mental reactions at the climacterium.**—G. R. JAMEISON and JAMES H. WALL. *Amer. Jour. Psychiat.*, 1932, **11**, 895.

THE authors' conclusion is that involutional melancholia is probably not a complete clinical entity in itself. The life history of the patient, including family tendencies, physical constitution, personality and psychic forces all have an influence. The period of the menopause with its physiological changes forms the background for the play of these several factors. Any type of borderline or frank mental disease may appear at this time. Four cases are briefly dealt with to illustrate some of the features that are considered most important.

C. S. R.

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[26] **A statistical study of hallucinations in the manic-depressive psychoses.**—KARL M. BOWMAN and ALICE F. RAYMOND. *Amer. Jour. Psychiat.* 1931, **11**, 299.

SOME 1,009 cases of manic-depressive psychoses were studied, with 1,408 cases of schizophrenia and 496 cases of general paresis as controls. It was found that hallucinations were absent in 70 per cent. of the males and 63 per cent. of the females in the manic-depressive group as compared with 25 per cent. of the males and 19 per cent. of the females in schizophrenia, and 73 per cent. of the males and 58 per cent. of the females in general paralysis. Auditory hallucinations were most common (16 per cent. males and 18 per cent. females). These were likewise most frequent in schizophrenia (50 per cent. males and 56 per cent. females). In general paralysis the incidence was nearly the same as in manic-depressive psychosis. Visual hallucinations were the next most common, while tactile, olfactory, gustatory, and somatic hallucinations were quite rare, there being less than 1 per cent. In schizophrenia the somatic types were most common (3 per cent. males and 4 per cent. females). Sex appears to play a rôle, as in all three psychoses females have more cases showing hallucinations. Martial condition and age seemed not to be related to the type or number, but there was some association between them and the prepsychotic personality. There was also considerable association between delusions and hallucinations, but the duration of an attack or the occurrence of a previous attack showed no relative factors.

C. S. R.

[27] **Encephalographic studies in schizophrenia.**—MATTHEW T. MOORE, DAVID NATHAN, ANNIE R. ELLIOTT, and CHARLES LAUBACH. *Amer. Jour. Psychiat.*, 1933, **12**, 801.

ENCEPHALOGRAPHY in a series of 60 selected cases of schizophrenia proved to be a safe clinical procedure. The cerebrospinal fluid pressures have, in the majority of cases, been normal or higher, indicating that the factor of a chronic increased intracranial pressure is present. The quantity of cerebrospinal fluid removed in the majority of cases has indicated varying degrees of cortical atrophy, and of enlargement of the ventricular system and cisternæ. The study of the encephalographic films has indicated a tendency to a selective atrophy involving the parietal lobes and the island of Reil area. The amount of mental deterioration is to some extent reflected in the degree of parietal lobe and island of Reil atrophy. Twenty-five cases showed evidence of increased size of the ventricles and cisternæ. None of the encephalographic films showed a normal cerebral pattern.

C. S. R.

[28] **Schizophrenia in epileptics** (Über Schizophrenie bei Epileptikern).—J. WYRSCH. *Schweiz. Atch. f. Neurol. u. Psychiat.*, 1933, **31**, 113.

THE coexistence of schizophrenia and epilepsy is rare but occasionally reported. In a series of 6,000 cases from a Continental institution it occurred eight times. That here reported concerns 10,000 admissions to another institution from 1873, when 16 examples of the combination, or of something analogous thereto, were found. Of these, several cases are cited in extenso. The conclusion is that in some there is independent development of the two states; in others, the schizophrenia has unusual characters and follows an exceptional course, being regarded preferably as an 'episode' of the epilepsy. By analogy, it then becomes comparable to the hallucinosis of malaria-treated general paralytics, to the psychoses of metencephalitis, to the schizophrenic syndromes of some organic cerebral processes, etc.

S. A. K. W.

[29] **The relation of the types of physique to the types of mental diseases.**—KATHERINE J. CAMPBELL. *Jour. Abnorm. Soc. Psychol.*, 1932, **27**, 147.

THIS study was made to verify or complete Kretschmer's generalization that certain types of physique are related to certain types of mental diseases. The 'pyknic' type, tending to be manic-depressive, and the 'asthenic' or 'athletic' types, tending to dementia præcox, were distinguished by the average weights and heights of each. When graphs and tables of distribution were made of these measures, male and female separately, and the manic-depressive and dementia præcox curves were laid side by side, the writer concluded that (1) all curves were fairly normal; (2) in four out of six comparisons, the curves of the manics and the præcox were practically the same; (3) the curve for weight in pounds showed that the male manics were on the average 10 lb. heavier than the male præcox; (4) the curve in pounds per inch of height showed that the mode weighed 0·10 lb. more per inch for the male manic, than for the male præcox. Kretschmer's difference was 0·30 lb. per inch.

C. S. R.

[30] **Age incidence of paresis.**—MAGNUS C. PETERSEN. *Amer. Jour. Psychiat.*, 1932, **12**, 521.

FROM the examination of the spinal fluid in 2,088 State Mental Hospital admissions, evidence of paresis was found in 179, or 8·2 per cent. A diagnosis of paresis was not made unless it was substantiated by definite serological tests. The youngest patient was a female of 22 years of age, and the oldest a

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male aged 77. Paresis was found more frequently among male than among female patients, except in the third decade when the ratio was almost reversed. The incidence curve of the male is sharper than that of the female and reaches its peak a decade later. Paresis of the aged, presumed by most text-book authors to be rare, was not infrequent in this series. The Wassermann reaction of the blood was negative in 18·5 per cent. of the paretic group as a whole, and in 30 per cent. of those 65 years of age and over. This undoubtedly explains why paresis of the aged often is diagnosed as arteriosclerotic or senile psychosis. Spinal fluid examination, which affords the only means of differentiation, is generally neglected in the absence of blood serological changes.

C. S. R.

[31] **Deletion of function and adaptive compromise.**—DONALD GREGG. *Amer. Jour. Psychiat.*, 1932, **12**, 511.

THE author concludes that 75 per cent. of diagnoses in the psychoses imply the deletion of function from organic causes. Eighty per cent. of symptomatic terms involve deletion of function. In studying the interpretation of the patient's behaviour (often interpreted in too subjective a manner) the rôle of deletion of function in producing such conduct is neglected, and we often forget the causes of such deletion of function wherein primarily lie hopes for amelioration and prevention.

C. S. R.

[32] **Measurement of mental deterioration in dementia præcox.**—RUDOLPH SCHWARZ. *Amer. Jour. Psychiat.*, 1932, **12**, 555.

THE Babcock test is here used. This test is based on the assumptions that the language ability of a deteriorating patient is the most resistive to the ravages of the disease, and that the difference between vocabulary and non-vocabulary tests indicates the degree of deterioration. In this study 110 dementia præcox patients showed a definite inferiority in their performance of a group of non-vocabulary tests compared with their ability shown in the vocabulary test. This inferiority may be expressed in percentages and was found to be 53·4 per cent. in the performance of the ' memory of paragraph ' test. It was noticed that with the longer duration of the disease and the deepening of the deteriorating process the size of the inefficiency index grew. This test, when found to be scientifically sound, may be profitably used as an auxiliary criterion to differentiate amentia from dementia, and benign mental processes from malign mental afflictions.

C. S. R.

## PSYCHOPATHOLOGY

[33] **Capillary changes in psychopathic children and young subjects following treatment with an iodothyroidin preparation** (Modificazioni dei capillari in fanciulli e giovani psicopatici trattati con un preparato jodo-tiroidinico).—G. PIGHINI and C. DELFINI. *Rassegna di studi psichiat.*, 1933, **22**, 3.

CAPILLAROSCOPY is considered by the writers of this article to disclose changes which are significant for advancing development when juvenile subjects affected by imbecility, idiocy, epilepsy, metencephalitis, cretinism, or mongolism are submitted to iodothyrin medication. Some 25 cases were thus treated, among which were 12 imbeciles or idiots. After a variable period, but usually some four or five months, observation of the capillary bed showed clearly that a previously existing ill-differentiated and -evolved capillary system was modified in the direction of greater evolution and distinction. The drawings which illustrate the change are rather impressive.

J. V.

[34] **The importance of the small blood vessels of the brain in psychiatric problems.**—N. W. WINKELMAN. *Amer. Jour. Psychiat.*, 1933, **12**, 775.

FROM a review of psychiatric work up to date Winkelman concludes that the histological processes responsible for mental abnormalities demand a study of the brain during the acute manifestations. The brain-changes in the various toxic and infective deliria tend to throw light on the problem of the mechanism of mental phenomena. Brain-changes in chronic processes, such as those due to lead, hypertension, and arteriosclerosis, will aid in obtaining a clearer concept of the physiology of the cortex. In order to evaluate completely the changes in the brain they must be studied in conjunction with the personality of the individual from whom it came. Just as different mental reactions can occur in dementia paralytica, depending on the basic personality of the individual, so in other mental diseases the same variability prevails and for the same reason.

C. S. R.

[35] **The sero-diagnosis of syphilis in mental hospital practice.**—J. ERNEST NICOLE and E. J. FITZGERALD. *Jour. of Ment. Sci.*, 1933, **79**, 52.

FROM prolonged experimentation the authors come to definite conclusions regarding the Wassermann reaction and the flocculations tests which are now well established as being two different ways of demonstrating the same phenomena. It is undoubted that the technique of the Wassermann test is cumbersome and almost out of reach of the smaller laboratories. The Was-

sermann test when compared with the flocculation tests shows the former to be very definite in sensitivity, while no more specific than some of the latter. Three flocculation tests may sound formidable, but they can be carried out in the average mental hospital with much more ease than any one Wassermann method alone. The question is the choice of flocculation tests. As an exclusion test which is easy, rapid, and capable of being applied to finger-blood, we have the micro-M.K.R. II and the Kline. If an exclusion *macro* test is required, the Kahn would appear most suitable. As the specific control, the routine Kahn seems second to none, and it can be used quantitatively. The third test could profitably be the M.B.R. II, a test that maintains a nice average between the sensitivity of the M.K.R. and the specificity of the Kahn. As regards spinal fluid, there seems less need for three tests, and here the Kahn and the M.B.R.Z. would seem eminently suitable.

C. S. R.

[36] **Mental phenomena observed in brain tumour.**—GEORGE W. HENRY. *Amer. Jour. Psychiat.*, 1932, **12**, 415.

NEOPLASMS in the brain according to their location give rise to mental phenomena by which the evolution of the cephalic portion of the nervous system may be traced. In the pontomedullary region mental phenomena are absent except for the late effects of intracranial hypertension. At times there is a disturbance of tactile sensibility, i.e., with one of the most primitive means of contact with the outside world. This disturbance is manifested by the appearance of tactile hallucinations. Those at the base show disturbances in growth and exquisite pain reactions. They give rise to automatic laughing and crying and to such crude motor phenomena as rigidity, tremor, choreo-athetoid movements, somnolence, stupor, drooling, automatic resistiveness, mutism, and in general to the manifestations of parkinsonism or catatonia. Occasionally the howling or other cries reminiscent of a lower mammalian stage in evolution are observed.

Cerebellar neoplasms may give rise to locomotor disorders, and the associated mental phenomena are about as crude as those of the pontomedullary region. Through increase of intracranial pressure lesions in the occipital lobe may produce marked visual illusory and hallucinatory phenomena. As the frontal lobes are approached the mental phenomena observed are increasingly complex. Lesions in the temporosphenoidal lobes, i.e., those affecting the archipallium, give rise to gross gustatory and olfactory disorders. In other words the response is a disturbance of primitive functions and of those senses upon which lower animals depend in their external contacts. Neoplasms of the temporal lobe may be accompanied by both auditory and visual hallucinations. These are usually highly organized and resemble the visions and voices of the functional psychoses. In the progression towards the frontal region visual hallucinations have been more

common. When the lobes are affected, however, highly organized auditory hallucinations are more frequently observed. Neoplasms in the frontal lobes also produce great personality changes and loss of the refinements of civilization.

C. S. R.

## PROGNOSIS AND TREATMENT

[37] **Diathermy in treatment of general paresis.**—HUGH A. MCKAY, KENNETH G. GRAY, and WILLIAM C. WINANS. *Amer. Jour. Psychiat.*, 1932, **12**, 531.

THE series presented in this paper comprised a total of 32 patients, and of this number 28 completed their course of treatment. Of these, three, or approximately 29 per cent., represent almost a complete remission; they have no physical defect, and no psychotic traits, nor outstanding mental defect; they are able to carry on trustworthy positions in hospital. Seven, or 25 per cent., represent partial remissions; they are able to carry on outside work under supervision. Three have physical defects, one has a memory defect, and one has considerable impairment of speech. Seven, or 25 per cent., are unimproved, but are quieter and cleaner in their habits. Three have died. All showed a marked physical improvement such as gain in weight and strength. Before treatment all showed a positive Wassermann reaction in the blood and spinal fluid, and a typical colloidal gold and colloidal mastic reaction. Afterwards there was found to be a change in the spinal fluid of nine patients. In six of these both the colloidal gold and mastic reactions became normal, but at the same time their Wassermann reaction remained unchanged. In one case the blood Wassermann test was negative and the fluid positive, with a normal colloidal gold reaction. The three remaining cases showed a favourable change in both the colloidal gold reaction and the colloidal mastic. Three of the cases with normal colloidal gold reactions were clinical remissions. In the other six cases the improvement in the serology did not parallel the clinical course.

C. S. R.

[38] **Sulphur pyretotherapy in mental diseases** (La sulforpiretoterapia nelle malattie mentali).—L. CERONI. *Rassegna di studi psichiat.*, 1933, **22**, 77.

SOME 50 cases representing various types of mental disease were treated by means of 'pyretogenic sulphur.' The results among the manic-depressives were particularly good, cures being claimed for a large proportion. In schizophrenia little improvement was obtained, and often none at all. The same remark applies to cases of epileptic insanity.

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[39] **Some remarks on the treatment of general paralysis by diathermy.**—NORMAN B. GRAHAM. *Jour. of Ment. Sci.*, 1933, **79**, 89.

IN 24 cases treated by the writer, 52 per cent. obtained a clinical remission. It is thought that the present results are superior to those following treatment by malaria. The mechanical control of the hyperpyrexia makes this method of treatment safe. Risk of burns is negligible if ordinary care is observed. Respiratory complications were a feature and caused one fatality.

C. S. R.

[40] **A review of seven years' malarial therapy in general paralysis.**—W. D. NICOL. *Jour. of Ment. Sci.*, 1932, **78**, 843.

Two hundred female cases were studied at Horton Hospital. The percentage of good remissions was low compared with the results of other observers. Leroy and Médakovitch state that in women the disease is less severe and tends to follow a more insidious course. They also report that the condition is more advanced before they become certified. The predominance of the dementing form in the female is probably responsible for the lower rate of remissions following treatment.

From the point of view of prognosis the writer lays most stress firstly on the clinical type assumed, and secondly on the duration of disease before treatment. His experience is that remissions are obtained by the first course of fever, and that subsequent courses of malaria exert no influence on the mental or physical progress. Benign tertian malaria is the most convenient species, but access should be available to quartan malaria for those patients who are immune to benign tertian, or who are in a poor state of health which might be endangered by the more severe type of fever. Whether second and further courses of malaria are necessary to maintain the latency or suppression of the syphilitic process cannot yet be decided.

C. S. R.

[41] **Therapeutic malarialization of general paralytics in the tropics.**—A. W. H. SMITH. *Jour. of Ment. Sci.*, 1933, **79**, 94.

GENERAL paralysis is as prevalent among the Malays as among the races of Europe. This prevalence is, moreover, even less than it would be otherwise on account of (1) the retarding effect of concurrent protozoal infections, and (2) the tendency of the Oriental's metabolism towards dermotropic rather than neurotropic lesions in all spirochætal diseases. General paralysis responds very much less to antimalarial treatment in Asia, because these neurotropic cases represent the most developed of the cases of lues, i.e. cases which have persistently resisted the action of exogenous and endogenous antibodies. It is extremely difficult to obtain effective malarial inoculation

in Asiatics of the coolie class who have been brought up in a country where malaria is indigenous, and who may be presumed to have acquired some degree of immunity.

C. S. R.

[42] **General paralysis : results of eight years of malarial therapy.**— B. Reid. *Jour. of Ment. Sci.*, 1932, **78**, 867.

From this study the author concludes that of all patients suffering from general paralysis, 31 per cent. are sent home improved each year. Six to nine years after treatment 12 per cent. of the patients are recovered from their illness and living at home. One to five years after treatment, 24 per cent. of the patients are recovered and living at home. Malarial treatment improves the physical condition of general paralytics even when no mental improvement results. As regards the expectation of life, 76 per cent. are living one year after treatment, 50 per cent. four years afterwards, and 22 per cent. nine years afterwards. Malarial treatment used on all cases of general paralysis admitted to hospital is not without danger, and 13 per cent. of cases die within one month of inoculation. The prognosis is best in those patients who have had symptoms of disease present for seven months or less prior to treatment. Reinoculation with malaria is of no special benefit in the treatment of general paralysis. This treatment also has little or no effect on the physical signs present in the nervous system.

C. S. R.

# Reviews and Notices of Books.

**Die Einteilung des Nervensystems nach seinen Leistungen.** By L. R. Müller, Professor of Internal Medicine, Erlangen. With 21 illustrations. Leipzig: G. Thieme. 1933. Pp. 74. Price M.6.80.

PROFESSOR MÜLLER makes in this short monograph a praiseworthy attempt to assign function to the different cell-fibre groupings of the central nervous system. His physiological conception divides the system into three main divisions, viz., (1) systema nervosum pro mundo, (2) systema nervorum myostaticum, and (3) systema nervorum vitale. As will be seen, these terms are equivalent to the more familiar 'voluntary' sensorimotor system, the automatic system of posture and equilibrium, and the vegetative nervous system respectively. The classification is doubtless somewhat schematic and scarcely indicates how they may severally be interrelated, but for descriptive purposes it has its value. Each of these is more closely analyzed, and, as remarked, an endeavour made to assign the functions comprised under each to specific mechanisms capable of localization. Much of course is well known and generally accepted, yet much more is still uncertain if not, indeed, frankly obscure. Economo's five types of cortical cell arrangement are specified, but association of function with the different cortical cell strata is at present, except on limited lines, virtually impossible. The different mechanisms of the myostatic system are, similarly, known in part, in part quite unknown. This portion of the subject is dealt with in a rather sketchy manner. As for the neurosympathetic system, little can be said to have emerged from numerous studies of recent years. The various ganglionic collections round the sides and floor of the third ventricle are all described and located anatomically, but present-day knowledge of their functions is still meagre. Some of the functions allotted by the author to some of the cell-fibre tracts are surely questionable; his view that the ansa lenticularis is connected with the mediation of bilateral emotional expression seems little else than guesswork, while to the corpus Luysii he assigns sympathetic functions concerned with the innervation of vessels and glands. There seems to be no allusion to an alleged Luysian syndrome of an entirely different kind.

The question of localization of psychical function is even more theoretical, but it is handled by the author in a very interesting way. A series of simple and well-drawn illustrations add to the usefulness of the essay.

**The Main Afferent Fibre Systems of the Cerebral Cortex in Primates.** By Stephen Poliak, M.D., Associate Professor of Neurology, University of Chicago. With 96 figures in the text. University of California Press, Berkeley, California. 1932. Pp. 370.

THIS is a very fine piece of anatomical research in monkeys, and raises questions of much interest and importance. Briefly, Professor Poliak has traced

to the cortex the three chief afferent systems of the body, viz., somato-sensory, auditory, and visual; he has fixed their cortical endings, and come to definite conclusions of which a few may here be annotated. As regards the first of these, the cortical termination of the main pathway of somatic sensibility extends more widely than was at one time supposed; thalamocortical radiation covers both sides of the rolandic fissure, and spreads far beyond, over the entire precentral agranular region, the whole of the post-central granular and part of the parietal agranular region; it also reaches to Brodmann's areas 1–6 and to 43. Further, the system is strictly unilateral, no fibres crossing by the corpus callosum. Dr. Poliak also shows that much more of the thalamus than its ventral nucleus is concerned with this somato-sensory pathway. The 'focus' of the cortical zone lies in the most anterior strip of the postcentral cortex at the foot of the rolandic sulcus.

As for the auditory radiation, it is proved to extend over the entire dorsal lip of the superior temporal convolution buried in the Sylvian fossa, and to part of the convexity of the same gyrus. The total area is smaller than might be for other reasons postulated, and perhaps represents only the minimum. Visual afferent fibres terminate exclusively in the area striata. The hypothesis of a retinal cortical projection beyond the area has no anatomical foundation.

In general, it may be said that each of these three afferent paths is a definite anatomical and physiological entity, distinct (though not absolutely separated anatomically) from other fibre-systems; the projection areas are gateways into the cortex, but the fundamental problems of how such zones are interconnected with other cortical fields and how 'higher processes' are mediated in relation thereto are still to be solved. Dr. Poliak's work approaches the conception of Flechsig, Henschen and other of the older investigators; his experiments substantiate the belief in representation of different functions in different cortical regions, and negative any theory of functional equivalence of the cortex as a whole.

S. A. K. W.

**The Tides of Life.** By R. G. Hoskins, Ph.D., M.D., Director of Neuro-endocrine Research, Harvard Medical School. London: Kegan Paul. 1933. Pp. 352. Price 15s. net.

UNDER a somewhat arresting if equally vague title Dr. Hoskins has compiled a general review of the whole endocrine situation as it exists to-day. To write a book which is at once popular (and therefore rather superficial) yet based on expert knowledge has always been regarded as a task of no little difficulty. The 'vulgarisateur,' when he succeeds, deserves well of his readers. In this particular case the rôle of presenting up-to-date scientific knowledge in pleasantly readable fashion has been well enacted, and we congratulate the author on his skill. For here the neurologist and endocrino-

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logist must not expect nice balancing of rival views or deep discussions of knotty points; yet there is no failure to cover the whole field, since the latest acquisitions—e.g., Cushing's pituitary basophilism—are duly described. So far as we have noticed, facts are carefully and accurately cited, inferences being drawn with commendable caution. The scientific spirit of the writer is well seen in his hesitation over the value of various endocrine preparations and his scepticism in regard to more than one. There are a number of good illustrations, but the bibliographical references are very incomplete.

**Ergot and Ergotism.** By George Barger, F.R.S., Professor of Chemistry in relation to Medicine in the University of Edinburgh. London and Edinburgh: Gurney and Jackson. 1932. Pp. 279. Price 15s. net.

THE interest of this monograph for the professional student of the nervous system resides mainly in its descriptions of ergotism from clinical and pathological standpoints. The author distinguishes gangrenous from convulsive ergotism, in this respect following what are now somewhat old if not actually obsolete descriptions. But the term 'convulsive' is misleading, and for it nervous or neural ergotism is much to be preferred. The nervous symptoms, we judge, are those of a toxic degeneration of peripheral and central neurones, so that they range from sensorimotor phenomena in the periphery to occasional fits and to dementia. The distinction made by the author from the gangrenous form is not valid, and he himself cites instances in which the two have been combined clinically. Antiquated accounts of symptoms (of which this book is full) are useless from the standpoint of pathology and pathogenesis; and there is no good reason to suppose that two separable forms or two different agents exist. On the contrary, ergotism resembles other known nervous affections in that either the periphery or the neuraxis may suffer. It is hardly necessary to point out that trifling lesions in the spinal cord will not explain the central phenomena of the condition, and that it is doubtful whether ergot convulsions can have a structural pathology. On the chemical and pharmacological sides knowledge is much farther advanced than on the clinicopathological. Recent Russian epidemics have brought pathological material, but little is said here of the actual findings. The Manchester epidemics of 1928 and 1929 are also referred to, but they were mild, and seemingly little if any pathological information was gleaned therefrom.

That deficiency in vitamin A may be a pathogenic factor is here alluded to as possible, on the basis of work by Mellanby; but much more knowledge of the objective facts of pathology is requisite ere the assumption can be validated.

It is much to be desired that the subject of nervous ergotism should be examined afresh by those who have expert knowledge of the nervous system, since it is still encumbered by abandoned conceptions. Part of the value of this truly fascinating monograph is, that it must stimulate interest in a

somewhat neglected chapter of nervous diseases, for text-books over the last half century have merely repeated uncritically old descriptions which are now out of date.

**Beitrage zur Kenntnis der Narkolepsie.** By Professor Dr. Rudolf Thiele and Priv.-Doz. Dr. Harmann N. Bernhardt, Berlin. Berlin: S. Karger. 1933. Pp. 187. Price M.18.

A SPECIALIZED study of narcolepsy was bound to make its appearance sooner or later, in view of the fact that cases have been occurring on an increasing scale of recent years and of the peculiar light which the phenomena throw on cerebral function. This essay deals with the subject along the usual lines of etiology, symptomatology, pathogenesis and therapy, and it covers the field with great thoroughness. The authors conclude (with Kinnier Wilson, Lhermitte, Redlich, and others) that narcolepsy cannot be considered a disease in the technical sense, a morbus sui generis, but rather a clinical syndrome of multiple causation. Nevertheless, they stress the admitted fact that some, perhaps many, cases are of endogenous nature, where some constitutional factor is at work, precipitating the affection, and they hold that these form a special class. The problem, it will be seen, is the same as that of epilepsy, in regard to which it is preferable to abandon the term 'idiopathic' and substitute 'cryptogenic.' Cryptogenic narcolepsy would signify that the cause is as yet undiscovered, but not that there is a narcoleptic disease distinct from the symptomatic variety.

The paragraphs on treatment refer to the value of ephedrin, and also, we are glad to note, to the usefulness of air injections—a recent development that promises well.

**Alcohol and Man.** Edited by Haven Emerson, M.D., Columbia University. New York: The Macmillan Company. 1932. Pp. 451. Price 18s. net.

A DISTINGUISHED editorial group of some seven writers is responsible for this volume, the latest contribution to a vexed problem. Addressed to the laity in the first instance (and therefore, presumably, containing a glossary in which it has appeared needful to define such words as 'alimentary,' 'sedative,' 'ovum,' 'hallucination' and others which we should have thought any educated person must know) the book discusses at some length such aspects of alcohol as its effects on normal man and experimental animals, its influence in varying dosage on personality and activity, its bearing on genetics and inheritance, therapeutic value, relation to longevity and life assurance, and kindred topics which need not be particularized. Much of the writing must of necessity be technical, and properly so, for the question of the action of alcohol on the systems of the body is technical, and cannot be 'popularized' without being rendered superficial. Accordingly, we find such terms employed as 'icteric,' 'endocardium,' 'urobilinogen' and scores of others of which no

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definition is furnished in the glossary, thus proving the unsatisfactory nature of any attempt to write for two classes of readers at one and the same time.

As a compendium of present-day knowledge this monograph can be consulted with much advantage. Its presentation of material derived from sources, in various instances, that are not readily accessible adds to its value as a work of reference; it contains numerous charts and tables in connexion with mental disease, death-rates, etc., and also selected bibliographies. A general critical spirit permeates the book, which is illustrated, for example, in the remark that the frequent claims made for the homosexual element in alcoholism need substantiation; the lowering of standards in the alcoholic environment and 'non-dependability of potency under ordinary stimulation' may well play a rôle.

**Methoden der Kriminalbiologie.** By Dr. F. v. Rohden, Alt-Scherbitz. Section No. 403 of the *Handbuch der biologischen Arbeitsmethoden*, edited by Geh. Med.-Rat Professor Dr. Emil Abderhalden. Berlin and Vienna: Urban and Schwarzenberg. 1933. Pp. 581–829. Price M.14.

In this section of Abderhalden's monumental handbook Dr. v. Rohden devotes more than 200 pages to a discussion of the methods of examination that should be adopted in the investigation of the criminal. With typical German thoroughness these are made to range over so wide a field in somatology, neurology, and psychology, that the task must consume weeks in a single case. Physical examination includes all the latest novelties, e.g., capillary microscopy and blood-group determination, also hunt for anomalies and stigmata of structure, etc.; study of the motor system comprises method of shaking hands, gesture and emotional expression, in addition to others either more, or less, familiar; graphology receives its due appreciation, also body build, physiognomy, and, on the purely psychopathological side, a large number of tests by which intelligence, character, personality and the like are, it is claimed, capable of being objectively assessed.

After analysis comes synthesis. The author describes various methods of classification. He cites statistics which are taken to prove that a high percentage of criminals, notably habitual criminals, are psychopaths; these he divides in different ways, e.g., impulsive psychopaths, psychopathic temperament types, psychopathic character types, complex psychopathic types. From another angle he discusses the criminals who are so from inclination, weakness, passion, conviction, and need, respectively. The view here taken is, that criminals are born and not made, in the sense that environmental factors are of much less pathogenic significance than those which are inherited or innate. How far the immense collection of data and the extraordinarily elaborate examinational methods here provided may be thought to contribute to a solution of the problem of the relationship of psychopathy to crime, is difficult to say. At least, they illustrate well its complexity.

**Nouveau Traité de Psychologie.** By Georges Dumas, Professor at the Sorbonne. Vol. II. Paris: Felix Alcan. 1933. Pp. 612. Price 100 fr.

THE second volume of the new treatise on psychology edited by Professor Dumas deals with the foundations of the life of the mind, hence include much that belongs in reality to physiology and biology. Indeed, the division of subjects is largely physiological; 'excitation and movement' is the general heading under which come discussions of cellular irritability, the neurone and the reflex, inhibition, conditioned reflexes and others classified as supra-elementary, tonus and still other motor phenomena. The secretions are next surveyed, and some account provided of how they are influenced by sensations, perceptions, and emotional states. Then follows a long and intricate study of sensation in its widest meaning which is very well done. A critical attitude is adopted towards the views of Head, although they are dismissed rather summarily and too little attention given to the matter of systemic duality. As might be expected in a work of the French school, anæsthesia receives due consideration. Next are examined the special senses. The third part of the volume is taken up with the emotional life and affective states; with pleasure and pain, their physiological basis and outward expression, and with the whole series of emotions and their psycho-physiological mechanisms. Desires, needs, and instincts are also studied seriatim. The James-Lange theory of the emotions is subjected to criticism along lines formulated by Cannon and others.

As a whole, this second volume maintains the high level set by the first, and forms a useful addition to the specialist's library. The reader might well have been spared, however, some unpleasant photographs of eastern provenance; aside from their intrinsic repulsiveness, they illustrate very badly indeed the particular point for which they have been inserted.

**The Nervous Child at School.** By H. C. Cameron, M.D., F.R.C.P., Physician in charge of the Children's Department, Guy's Hospital. London: Oxford University Press. 1933. Pp. 160. Price 6s. net.

DR. CAMERON'S thoughtful, sensible, and encouraging little book on the nervous child at school should be read from cover to cover not only by distressed parents and pædogogues but by the neurologist and psychiatrist as well as the practitioner. In simple language, free from technicalities, the problems offered in these days by sensitive children at preparatory and by young people at boarding schools are first defined and then faced. Broadly speaking, Dr. Cameron finds in fatigue the cause of unhappiness in school life, and offers many wise counsels as to the best ways of combating it. He outlines clearly the vicious circles which arise only too readily as parental disappointment combines with school failure to intensify in the mind of the nervous child his difference from his associates and his longing to escape. Disorders of health and conduct incidental to this unhappiness or distinct

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from it are examined concisely and their treatment sketched. Dr. Cameron possesses a happy insight into the heart of youth and is able to put himself in the place of the sensitive boy so sympathetically as to gain inside knowledge, so to speak, whereby to solve the knotty questions that turn up every now and again. We are glad to note that the appeal of the book is not limited to those whose circumstances permit education at preparatory or boarding schools; the author's remarks apply in full force to life at day schools also. There is throughout a gratifying absence of stress on sexual factors and freedom from the fads of the day in respect of early sex information and other things; Dr. Cameron is dealing not with defectives and delinquents and frankly abnormal beings but with children who are not outside normal limits. He admits the powerful factor of sex in the life of every boy and girl, yet insists convincingly that the cases with which he deals are but indirectly concerned with any such element. This sex-free aspect of the question is underlined all too seldom in these days. He has more than a word for the unfortunate mental attitudes of anxious parents and, indeed, of some irritated masters. The book can be put with confidence into the hands of all who have to deal with growing youth.

**Diseases of the Spinal Cord.** By Williams B. Cadwalader, M.D., Professor of Clinical Neurology, University of Pennsylvania Medical School. London: Baillière, Tindall and Cox. 1932. Pp. 204. Price 29s.

A SCHEMATIC division of diseases of the nervous system in accordance with their anatomical site is becoming, we are inclined to think, rather obsolete. Better acquaintance with the processes of nervous disease throws doubt on the strict localization of such states to this or that section of the neuraxis. This is certainly true of peripheral and of central infections, and is supported by evidence derived from encephalitis, meningitis and other generic conditions. We do not speak much of diseases of the cerebellum, for the simple reason that that organ is so related to other neural structures as to render limited cerebellar disease a rarity. The remark applies also to the spinal cord, and Dr. Cadwalader is naturally well aware of this. He includes in his book 'syphilis of the spinal cord,' and, if so, why not all varieties of spinal meningitis? Arachnoiditis circumscripta is mentioned, but not, seemingly, pachymeningitis hypertrophicans; suppurative extradural meningitis or abscess is described, but not intraspinal abscess. Conversely, we find included both the muscular dystrophies and Thomsen's disease, though their connexion with the cord is remote; nor can the writer discuss tabes without alluding to its ocular manifestations, or syringomyelia without reference to syringobulbia. Hæmatomyelia is dismissed in a single short sentence, and thrombomyelia likewise. There appears to be no notice of spinal embolisms. Such comments are not made with any other end in view than to stress the unsatisfactory nature of any effort at placing spinal maladies in a group by themselves.

So far as the author's descriptions are concerned, the book serves its

purpose well; they are clearly set forth and illustrated. The chapter on topographical diagnosis contains useful matter, as do the initial sections on anatomy and physiology. There is a good bibliography to complete the work.

**Human Values in Psychological Medicine.** By C. P. Blacker, M.D., M.R.C.P. London: Oxford University Press. 1933. Pp. 179. Price 8s. 6d. net.

THIS attractively written and thoughtfully conceived book consists in what might be termed a series of essays, not always closely related in any superficially evident way, which deal with differing aspects of the mental life, normal and abnormal. Chief attention is engaged by questions of 'values'—the meaning of the word, its uses, and its applications. The word, as noun or verb, has both intellectual and affective connotation; values are as varied and manifold as psychic needs. Distinctions can be drawn between the commercial, biological, and sentimental value of a thing. Elaboration of the subject takes the author next to the matter of judgment values, æsthetic values, pivotal values ('those which, in one way or another, unify and justify life, give it coherence and make it on the whole worth living'). Discussion of psychopathological patients' values follows. Much is said also of religious values, and of attitudes of mind in connexion therewith. The author then passes to consideration of 'realities,' which word in his view has four distinct and separate meanings. He argues that the difference between his 'physiogenic' and 'psychogenic' reality is the same as that between 'functional' and 'organic' in neurology. A closing chapter deals more or less on Freudian lines with life and death instincts, though the author's own hypothesis is in a sense distinct from the Freudian.

**The Sign of Babinski.** By John F. Fulton, Professor of Physiology in the Yale University School of Medicine, and Allen D. Keller, Professor of Physiology in the School of Medicine, University of Alabama. London: Baillière, Tindall and Cox. 1933. Pp. 165. Price 26s. 6d. net.

THE theme of this mainly physiological study is the evolution of cortical dominance in primates; and in a sense only a minor part of the work is taken up with the subject which gives it its name. The authors have made a praiseworthy attempt to determine the correlates of clinical reflexes in the higher apes, and from this standpoint the book has its value. Its real appeal, no doubt, is to the neurophysiologist, concerned as it is with extirpations of parts of the motor cortex in the higher apes (gibbon, chimpanzee, baboon, etc.) and with study of the resultant phenomena. The clinical examination of the Babinski response and the discussion of its significance, here offered, will hardly satisfy the requirements of the neurologist. The authors have thought it well to reproduce on an almost microscopic scale the original article by Babinski, which appeared in the *Semaine médicale* in 1898. Why they should have considered this desirable, or assumed that the journal is

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'now difficult of access,' does not appear; it can be found in any medical library worthy of the name.

**A New Approach to Dietetic Therapy.** By Eugene G. Földes, M.D., formerly Assistant Professor of Medicine, Budapest. Boston: Richard G. Badger. 1933. Pp. 434. Price $5.00 net.

THE part of Dr. Földes' book that has neurological interest is that in which the relation of the phenomena of epilepsy and migraine to disorders of water and mineral metabolism is examined. Aside from visible water retentions, such as the œdemas, Dr. Földes is at pains to emphasize the clinical and pathogenic import of non-visible retentions, and also of liquid (water plus mineral) immobilization. Recent studies have shown the great part played by such disturbances in connexion with epileptic attacks, and this aspect of the question is fully gone into; what Dr. Földes does not make clear is whether he regards the retention of water as the sole, or only one, factor precipitating seizures. From what he says on p. 67 ('the development of epileptic attacks is due to a temporary accumulation of liquids in the tissues and spaces enclosed by the capsules of the brain, thus leading to increased pressure in these tissues and spaces and thus to irritation of cortical centres') it would appear that he takes the water factor to be self-sufficient—indeed, he states elsewhere that it 'is a basis upon which all observations can be understood from the standpoint of a single entity.' We fear this claim is ill-founded; while the importance of the water-retention factor is not to be gainsaid there are varieties of epilepsy in which no good reason to postulate its presence in action can be advanced. Further, by making it pathogenically unique injustice is done to other precipitants whose coexistence cannot be denied. In a word, we consider that Dr. Földes here overstates his case, and this is the more regrettable as we are equally convinced that his work is of much value and that he has proved his general thesis. The applicability of the water-mineral disorder theory to other morbid states here dealt with must be allowed.

**Case Studies in the Psychopathology of Crime.** By Ben Karpman, M.D., Professor of Psychiatry, School of Medicine, Howard University, Washington, D.C. Washington: Mimeoform Press. 1933. Pp. 1042.

THIS big quarto volume of over 1,000 pages contains long accounts of the investigation of five criminal cases in which there was good reason to suppose psychopathological features were too pronounced to be ignored. The first was diagnosed, apparently, as one in which the subject's reactions were considered as 'an overcompensation for a deep sense of defeat.' In the second, the man was held to belong to 'that class of drifters whose life is one long childhood,' and who get into crime 'as a matter of course'; emotional and mental sides of such beings do not keep pace. The third was a homosexual. As for the fourth, he had a 'hysterical make-up' and might have been spared

a criminal career had circumstances not been too much for him. The fifth lived more 'in phantasy than reality,' was constitutionally 'inferior,' and though able to distinguish intellectually between right and wrong, was unable to choose the right thing 'emotionally.'

We have nothing but praise for the infinitely painstaking fashion in which Dr. Karpman has collected his material and for his laborious effort to present it in consecutive, logical, and readable manner. Yet we confess to a certain disappointment, inasmuch as the autobiographies of the criminals bulk far too largely in the volume and become positively tedious; they are filled with trivialities of no significance whatsoever. We are given life-histories which might easily have been blue-pencilled without detriment; by being rendered more concise they would have been more impressive. Moreover, the student of the complex problem of the interrelation of crime and personality would have welcomed more technical discussion of the bearing of the selected cases on that question. Incidentally, the light thrown on the penitentiary conditions of certain American institutions is extraordinarily revealing.

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